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Date 08-27-2003
ABSTRACT

Adolescence is a time when individuals begin to explore and examine psychological characteristics of the self in order to discover who they really are and how they fit in the social world in which they live. It is during this time of self-exploration that adolescents at risk for Huntington's Disease often learn of their risk status and witness the debilitating symptoms of the disease in their parents. Huntington Disease (HD) is an autosomal dominant neuropsychiatric disorder characterized by mid-life onset, involuntary movements, cognitive impairment, and depression.

This dissertation investigated how adolescents experience living in a family with Huntington's Disease and therefore at risk for Huntington's Disease, and how this impacts their self-understanding and self-identity. The method of inquiry was based on a phenomenological approach. In-depth interviews were conducted with each of the adolescents. The data were analyzed using Van Manen's (1980) and Cochran and Claspell's (1987) format, resulting in an extraction of three themes. These themes are: (1) Naming the Legacy: Understanding and Misunderstanding; (2) Experiencing the Legacy: Huntington's Disease in Relation to Relationships; and (3) Integrating the Legacy: At the Crossroads of Self and Future Self.

The analysis emphasizes that the at-risk adolescents' exploration of self-identity and future self was an individual process influenced by the cognitive, developmental, and socio-cultural contexts of the adolescents' lives. The process of learning about Huntington's Disease occurred through intuition and practical and experiential learning. The adolescents found support outside their family through friends and adult mentors. They engaged in complicated coping strategies and demonstrated a capacity for decision-making that displayed maturity beyond what would be expected for their age group. These findings led to specific recommendations for theory, research, and clinical practice in the area of the adolescent experience of HD. The
research underscores the need for healthcare professionals to re-evaluate their view of adolescent autonomy and capacity for decision-making.
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ACKNOWLEDGEMENTS

This thesis evolved from conversations with one of my clients ten years ago. She sat in my office and asked me if I knew about Huntington’s Disease. I confessed I knew little about the disease. She told me her story of being at risk and living in a family with HD. She asked me to support her as she went through the process of predictive testing. Little did I know that on that day I would begin a journey into the uncharted territories of genetic disease and predictive testing that finds me today attempting to describe the experiences of adolescents at risk for HD.

As my journey began, I went to seek more knowledge about the disease from Susan Tolley, the Director of the Huntington Disease Resource Centre. Susan not only provided me with information, she graced my journey with her wisdom, support, and perspective. Few travelers can begin a journey with the aid of such wise counsel.

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DEDICATION

For my Grandfather

Jesse Leroy Bloch (1904-1959)
CHAPTER I
INTRODUCTION

Part of the fascination of the new genetics concerns the questions it raises about the construction of knowledge - how, for whom, and for what is this knowledge being constructed? In this context, then, Huntington’s Disease may serve as a space where many discourses collide and therefore help make visible the hidden stakes in this contest for human survival and identity in which all of us are at risk. Alice Wexler (1995:xxv)

This thesis is primarily a use of the qualitative method of phenomenology, enriching the practices of counselling and clinical genetics, with a focus on the life experiences of adolescents at risk for Huntington’s Disease (HD). It was initially motivated by ethical concerns but it does not explicitly address the ethical implications or engage in “second-order” reflection on ethical theory. In particular, I am interested in how adolescents experience living in a family with Huntington’s Disease and, therefore, at risk for Huntington’s Disease, and how this impacts their self-understanding and self-identity.

Adolescence is a time when individuals begin to explore and examine psychological characteristics of the self in order to discover who they really are and how they fit in the social world in which they live. It is during this time of self-exploration that adolescents at risk for Huntington’s Disease often learn of their risk status and witness the debilitating symptoms of the disease in their parents.

Although many issues exist in any family group with adolescent-age children, additional dilemmas confront those families living with a chronic and debilitating illness like Huntington’s Disease. And for children of Huntington’s Disease-affected parents the challenges are daunting. Unfortunately, there is very little information or research regarding the experience of the at-risk adolescent living in a family with Huntington’s Disease. As a result, there are few interventions tailored to help adolescents deal with the challenges that the disease presents. It is the intention of this research to address the needs of the adolescent at risk for Huntington’s Disease by asking,
What is the experience of adolescents at risk for Huntington’s Disease, and how does this experience impact self-understanding and self-identity?

Adolescence

Adolescence is a transitional period in the human life span – linking childhood and adulthood – that involves biological, cognitive, social, and emotional changes. Cultural and historical circumstances limit our ability to place a definitive age range on adolescence; however, in North America it is generally agreed that adolescence begins at approximately thirteen years of age and ends, for most individuals, between the ages of eighteen and twenty-two (Compas et al., 1995). Adolescence is described in terms of early and late periods. Early adolescence corresponds roughly to middle or junior high school years and includes most pubertal change. Late adolescence refers to the latter half of the second decade of life. Career interests, dating, and identity exploration are often more pronounced in late adolescence than in early adolescence (Cormack, 1992).

The biological, cognitive, social, and emotional changes of adolescence range from the development of sexual functions to abstract thinking processes and independence. During adolescence, individuals are beginning to understand themselves and to develop a self-identity (Erikson, 1968).

Adolescence is also characterized by changing and evolving relationships both inside and outside of the family (Furman & Wehner, 1993). While establishing the boundaries of their own identity, young people seek validation from their peer group (Piaget, 1932; Erikson, 1968). It is during this complex time of self-understanding and identity formation that adolescents may learn that a family member has Huntington’s Disease and that they in turn are at risk for the disease.
Huntington’s Disease

Huntington’s Disease is an inherited degenerative neuropsychiatric disorder that is characterized by involuntary movements, profound personality change, cognitive impairment, and depression. Onset of the disease typically occurs in mid-life (mean age of onset is thirty-eight), although much earlier and later occurrences are also known (Hayden, 1981). At present there is no effective treatment or cure, and ameliorative actions are largely restricted to the administration of anti-depressant drugs (Ranen et al., 1993). The progression of the disease is gradual but inexorable, and death usually occurs fifteen to twenty years after onset. In general, the patient dies not from the disease itself but from complications such as pneumonia, heart failure, or infections resulting from the weakened condition of the body. Because of its typical late onset and devastating progression, the disease is described as a “genetic time bomb” that “remains dormant until the person reaches adulthood” (Huntington’s Disease Society, 1994).

Huntington’s Disease occurs in approximately 1 in 10,000 of the general population (Hayden, 1981). For every affected person there are approximately five to seven persons at risk, either as children (50% risk) or grandchildren (25% risk). As an autosomal disorder, HD affects both men and women, and either the father or the mother can pass the gene on to the next generation. Huntington’s Disease is also a dominant disorder, which means that all offspring of an individual who carries the gene for HD will have, at birth, a 50:50 chance of either inheriting or escaping the disease. Inheriting the gene for Huntington’s Disease is, therefore, a random event that Wexler (1992) compares to the flip of a coin. Not inheriting the gene (an equally chance event) means the “chain is broken,” and HD will not reappear in subsequent generations. The mutation is highly penetrant; thus anyone who inherits it will, if they live long enough, “almost certainly” become affected (Cox & McKellin, 1999:623).
In 1993, the identification of a novel gene containing a trinucleotide repeat provided the opportunity for direct mutation testing of an individual blood sample (Huntington’s Disease Collaborative Research Group, 1993). This trinucleotide sequence of CAG repeats from eleven to thirty-four in normal chromosomes, and from forty-two to over sixty-six in HD chromosomes (Drlica, 1994). This region of repeats seems to be unstable in the disease form of the gene, with higher numbers of repeats correlating with earlier onset of disease symptoms. Exactly how a high number of CAG repeats leads to disease is not yet known; however, its correlation with disease should make the test for HD much more accurate in the future (Duyao et al., 1993; Simpson et al., 1992).

When a family member receives a diagnosis of Huntington’s Disease it is inevitable that changes in family communication will result and create the possibility of family members learning for the first time of their risk status (Yale and Martindale, 1984). The knowledge that HD runs in the family creates for those at risk a state of prolonged and anxious uncertainty (Barette & Marsden, 1979). The impact of this information on at-risk children and adolescents has not yet been the subject of detailed study.

**Predictive Testing**

The “genetic revolution,” fuelled by the Human Genome Project, is engaged in decoding the messages contained in human DNA. The Human Genome Project has mapped the genome and identified approximately 30,000 genes (Collins et al., 2003). Once a gene is located and its sequence of chemical code letters discerned, scientists can devise a test for the presence of that gene in any patient. Tests for the presence of genes that either cause disease or make people more susceptible to disease are becoming increasingly available. In some cases, such knowledge may lead to treatments that delay the onset of the disease or soften its effects. In other instances, almost nothing can be done to stop the severe effects of disease brought on by genetic mutations.
The evolution of genetic technology and the information it provides through diagnostic and predictive testing has created complex social, ethical, and legal concerns. These concerns arise not only from the information that this technology provides about individuals, but also from the impact of the results of this knowledge upon individuals and families.

In the field of predictive genetic testing, technology has advanced far more rapidly than our understanding of its larger social significance and consequences. In what way will this information affect our understanding of health, illness, and our relationship with others? According to Kenen (1996:1549), “Accepting the at-risk health role involves complex psycho-social relationships with family members, particularly with those who will become caretakers, if that becomes necessary, and interpersonal relationships, in general, can be particularly problematic when a genetic disease runs in the family.” How will individuals and families at risk for inheritable disorders come to understand the information these tests provide? Wexler (1992) cautions that people can misinterpret genetic information and the implications of such tests. Moreover, the information often reaffirms risk and offers no cure. Adolescents at risk for Huntington’s Disease can learn of a parent’s positive test result and learn decisively that they are at risk for the disease. How would growing up at risk for HD affect an adolescent’s development and self-understanding within the family and in relationships outside of the family?

It is the ability to predict future disease that separates predictive testing from most other biomedical or diagnostic testing, and this ability brings with it information that affects not only the patient but the family. As Cox points out, “By virtue of the fact that we inherit DNA from both parents and share an average of 50% of the same genes as our siblings, genetic information does not ‘belong’ to just one person” (1995:4). Who does this information belong to?

What is the appropriate practice for disclosure when the issue of confidentiality and the perceived responsibilities to the at-risk individual’s family are in the balance? Wertz and Fletcher (1989) conducted an international study of medical geneticists and found that only one-
third of the 700 respondents would endorse the privacy of the individual patient with Huntington’s Disease, while another one-third would tell the patient’s relatives. As the study demonstrates, regulated professionals have differing ideas about disclosure.

Outside of the clinical setting how do families begin to understand genetic information and manage in their everyday lives? Yale and Martindale (1984) argue that it is essential to consider the needs of the entire family at the time of diagnosis of a family member, as this may precipitate new discussions of genetic risk that could lead to family members discovering for the first time that they are at risk for HD. Folstein (1989) argues that adults who learn about their risk status are often completely preoccupied with this information, that children may be less disturbed because developmentally they are not able to understand the implications of their risk status, and that adolescents may be shielded by the feeling of invulnerability that teenagers experience at this developmental stage. Age-specific responses, however, have not been the subject of detailed analysis. Moreover, the clinical observations of Folstein (1989) and others may underestimate the potential adverse responses of children and adolescents to the information that a parent has the gene for HD and that they are at risk for HD.

**Familial and Social Dynamics**

Within the medical genetics community, predictive testing for Huntington’s Disease has provided a paradigm for understanding many of the difficult problems that are inherent in testing for adult-onset disorders. Adding to the complexity of Huntington’s Disease is the fact that the onset of symptoms in adults often coincides with an already challenging stage of life – the parenting of adolescent children.¹ For adolescents, it is often during the complex time of identity

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¹The typical age of onset is between thirty-six and forty-five years but the disease may remain quiescent until the individual is well into old age. At the opposite end of the life-course, the juvenile form of HD may manifest itself during adolescence or, in rare cases, in early childhood (Hayden, 1981).
formation that they learn that a family member has the disease and that they in turn are at risk for the disease. Current research is beginning to explore the psychological effects of predictive testing on at-risk individuals and family members (Tibben et al., 1993a; 1993b; 1993c; Cordori & Brandt, 1994a; 1994b). There has been little focus on how Huntington’s Disease affects the familial and social dynamics of affected persons and their family members. The response of children and adolescents to learning about their risk for HD, and the impact of this risk status on their lives, has not been the subject of detailed analysis.

There is an urgent need to know more about how growing up in a family with a history of genetic disease shapes an individual’s self-identity and relationships with others, both inside and outside of the family (Richards, 1996). It is the intention of this research to investigate and illuminate how adolescents experience the presence of Huntington’s Disease in the family and of being at risk for the disease. In particular, I am interested in how adolescents learn about HD, the impact of this knowledge on relationships with family and friends, and on self-understanding, self-identity, and future-self.

Summary of Chapters

A brief summary of each chapter in the dissertation follows:

Chapter II will explore the history of adolescence. In order to provide the background necessary to explore the impact of HD on the self-understanding and self-identity of the at-risk adolescent, the following theories on adolescent development will be reviewed: Piaget’s (1967) cognitive developmental stages, Harter’s (1990a; 1990b) dimensions of self-understanding, and Erikson’s (1968) theory of self. The chapter will conclude with a review of the literature on adolescence and Huntington’s Disease.
Chapter III provides an overview of the phenomenological research method. The methodology is reviewed and the impact on the choice of methodology to the research design is discussed.

Chapter IV presents a discussion of the co-participants' lifelines – a chronological timeline that begins with the adolescents’ first intuition of Huntington’s Disease – and the stories that developed through the interview process. Marilyn’s story will be used as an example of the adolescent experience of living in a family with HD and at risk for HD. Her story was chosen because it reflects the themes that emerged across all the adolescents’ narratives. Marilyn’s story will present and illustrate the three themes that emerged from the stories and that characterize the experiences of adolescents in families with HD: (1) Naming the Legacy: Understanding and Misunderstanding; (2) Experiencing the Legacy: Huntington’s Disease in Relation to Relationships; and (3) Integrating the Legacy: At the Crossroads of Self and Future Self.

Chapter V, Naming the Legacy, describes three stages in the process of learning about Huntington’s Disease. The chapter begins with an analysis of the adolescents’ intuition that “something is wrong.” The next section of the chapter explores “who” told the adolescents about HD, and illuminates “how” the adolescents learned about HD and “what” they learned about HD. All of the adolescents demonstrated two types of learning: practical learning and experiential learning. Practical learning involved learning from an adult that a parent had a disease called HD. Experiential learning occurred when the adolescents began to experience their parent’s symptoms of HD and to distinguish those symptoms from other behaviour. In the last section there will be an exploration of what the adolescents understand about HD, what they tell their friends, and their experience of family secrets and misunderstandings.

In Chapter VI, Experiencing the Legacy, the effects of Huntington’s Disease on family and peer relationships will be explored. Family relationships will include the parent/child
relationship, sibling relationships, and extended family relationships. Non-family relationships will focus on peers.

In Chapter VII, Integrating the Legacy, the dimensions of self-understanding will be examined to explicate the complex process of identity formation and to explore the impact that being in a family with HD has on identity development. It will be demonstrated that the experience of the adolescent at risk for HD is best characterized as an individual process that is influenced by cognitive and developmental levels and the socio-cultural context of the adolescent’s life. The narratives of the adolescents who participated in this research will be used to illustrate these dimensions and to establish the role that Huntington’s Disease plays in familial and social experiences and how these experiences contribute to self-understanding and understanding of future self.

The second section of the chapter will examine the adolescents’ views on their future, personal meaning of at-risk status, and their views on pre-symptomatic testing. Chapter VIII will provide a comprehensive overview of the research. The three themes that emerged from the interviews will be explored in such a way as to illuminate the experience of at-risk adolescents and the impact of these experiences on self-understanding and self-identity. The chapter will also include an examination of the coping strategies employed by the adolescents living at risk in families with HD. These coping techniques involved personal, interpersonal, and group behaviour.

Chapter IX will discuss the implications of this study for theory, future research, and practice.
CHAPTER II
LITERATURE REVIEW

Adolescence is a border between adulthood and childhood, and as such it has a richness and diversity unmatched by any other life stage. . . . Adolescents are travelers, far from home with no native land, neither children nor adults. They are jet seters who fly from one country to another with amazing speed. Sometimes they are four years old, an hour later they are twenty-five. They don't really fit anywhere. There's a yearning for place, a search for solid ground. (Pipher, 1994:74)

For much of the twentieth century, Western culture has perceived adolescence as a problematic period in the human life span. Beginning with G. Stanley Hall's (1904) portrayal of adolescence as a period of "storm and stress," ideas and attitudes about adolescence have tended to stereotype and marginalize youth, and often reflect little of the actual adolescent culture.

Purpose and Outline of Chapter

This chapter will explore the history of adolescence. A review of Piaget's (1967) cognitive stages of development, Harter's (1990a, 1990b) dimensions of self-understanding, and Erikson's (1968) stages of self-identity will be elucidated to provide the reader with background information on the major theories on adolescent identity development. The chapter will conclude with a review of the literature on adolescents and Huntington's Disease and the development of predictive testing.

Summary of Research on Adolescence

Plato to Rousseau

Adolescence is a transitional period in the human life span, linking childhood and adulthood. It is not necessarily a stage that human beings pass through but rather a segment of continuing human development. While adolescence has some unique characteristics, what takes
place in adolescence is interconnected with development and experiences in childhood and adulthood.

The contemporary Western idea of adolescence as a period marked by conflict and disturbances is largely a creation of the late nineteenth and early twentieth century. The transition from childhood to adulthood has been recognized as significant since the Greeks. Plato argued that reason emerges in youth, and that childhood experiences influence the period of time we now call adolescence (Plato, fourth century B.C./1968 translation). Aristotle believed that the ability to choose was an important aspect of young manhood, and that self-determination was the hallmark of maturity at this stage. Aristotle was one of the first individuals to describe specific time periods for stages of human development: "boyhood," for example, was understood to cover the period from age seven to puberty, and "young manhood" from puberty to age twenty-one (Aristotle, fourth century B.C./1941 translation). Such views are not unlike contemporary ones, which use labels like independence, identity, and career choice to ascribe the importance of increased self-determination to the period of adolescence (Amundson and Borgen, 1995).

During the Middle Ages children were not given a status apart from the adult and, consequently, they were treated as if they held the same interests as adults (Muuss, 1989). In the eighteenth century, Rousseau described a more enlightened view of adolescence (Emile, 1762/1962 translation). His views were similar to Aristotle and Plato in that he believed that development in childhood and adolescence occurs in a series of stages. He argued that reason and self-consciousness developed at twelve to fifteen years of age, and that emotional maturity replaced selfishness at fifteen to twenty years of age.

The Age of Adolescence

In 1904, G. Stanley Hall published Adolescence, a two-volume work that played a major role in restructuring thinking about adolescence, and that initiated the scientific study of
adolescent development. Hall applied Charles Darwin's theory of evolution to the study of adolescent development. He believed that all development was controlled by genetically determined physiological factors, and that environment played a minimal role in development, especially during infancy and childhood. He did acknowledge, however, that environment accounted for more change in development during adolescence than during other periods. Hall believed that heredity interacted with environmental influences during this period to determine an individual’s development.

According to Hall, adolescence was the period from twelve to twenty-three years of age, and was filled with “storm and stress” (Hall, 1904). This storm and stress period defined a turbulent time charged with conflict and mood swings. Adolescents’ thoughts, feelings, and actions oscillated between conceit and humility, good and temptation, happiness and sadness. Hall argued that biological changes in adolescence allowed for more complicated social arrangements, such as dating. Hall also argued that after the age of fifteen emphasis should be placed on education in areas such as civility, scientific thinking, and morality.

In 1928, Margaret Mead studied adolescents on the South Sea island of Samoa (Mead, 1928). She concluded that adolescence is most importantly affected by sociocultural factors not biological ones. She argued that when cultures provide a smooth, gradual transition from childhood to adulthood, little “storm and stress” is associated with the period. Mead’s observations of Samoan adolescents revealed that their lives were relatively free of conflict. Mead concluded that cultures that allowed adolescents to observe sexual relations, see babies born, regard death as natural, do important work, engage in sex play, and clearly observe adult roles promoted a relatively stress-free adolescence. In cultures like the United States, in which children are considered very different from adults, and where adolescence is not characterized by the aforementioned experiences, adolescence is more likely to be stressful. Mead was the first to
point out the wide variability in adolescent adjustment, and the contribution of social and cultural factors associated with the period.

While adolescence has a biological base, as G. Stanley Hall believed, it also has a sociocultural base, as Margaret Mead showed. Additionally, sociohistorical conditions have contributed to the emergence of the concept of adolescence. The inventionist view contends that adolescence is a sociohistorical creation (Lapsley, Enright, & Serlin, 1985) that was “invented” in response to the movement to create a system of compulsory public education. Especially important to the inventionist view of adolescence were the sociohistorical circumstances at the beginning of the twentieth century, a time when legislation was enacted that ensured the dependency of youth and made their move into the economic sphere more manageable (Mirel, 1991). Some of these notable sociohistorical circumstances in North America include the decline in apprenticeships, increased mechanization as a result of the Industrial Revolution (which also required the development of new skills and specialized divisions of labour), the separation of work and home, increased urbanization, age-segregated schools, and the establishment of youth groups such as the YMCA and the Boy Scouts.

Between 1890 and 1920 a great deal of compulsory legislation in North America aimed at youth was enacted. Tyack (1976) notes that two clear changes for youth resulted from such legislation: decreased employment and increased school attendance. In the United States from 1910 to 1930, the number of ten- to fifteen-year-old adolescents who were gainfully employed dropped by about 75%. In addition, between 1900 and 1930 the number of secondary school graduates substantially increased. Approximately 600% more individuals graduated from secondary schools in 1930 than in 1900.

By the turn of the twentieth century, most adolescents in North America lived with parents, were economically dependent on parents, had few or no responsibilities outside of school, and were slowly acquiring the power to choose their own occupation and mate.
Eventually, multiple options regarding occupation and marriage came to characterize the adolescent experience. In short, the adult identity came to be a matter of choice and definition for adolescents themselves. This was paralleled by changes in the structure and social psychology of the family: The family no longer constituted a self-sufficient economic unit. It was made up of fewer children who tended to be closer in age, and the average age of home-leaving increased (Baumeister, 1991).

Erikson (1968) contends that structural developments in society, as well as changes in the experiences of adolescents, contribute to the creation of a “psychological moratorium” — a term Erikson applies to the gap between childhood security and adult autonomy, which adolescents experience as part of their identity exploration. He claims that during adolescence individual’s experience “identity confusion.” Identity versus identity confusion constitutes one of eight developmental stages outlined by Erikson.² Erikson contends that during this time adolescents face finding out who they are, what they are all about, and where they are going in life. Adolescents are confronted with the possibility of many new roles, such as vocational and romantic roles. As adolescents explore and search their culture’s identity files, they often experiment with different roles. Erikson believes that adolescents face an overwhelming number of choices, and at some point during youth enter this moratorium. They try out different roles and personalities before they reach a stable sense of self. They may be argumentative one moment, cooperative the next. They may like a particular friend one week, despise the friend the next week. Erikson contends that this personality experimentation is a deliberate effort on the part of adolescents to find out where they fit in the world.

² In Erikson’s theory, eight stages of development unfold as we go through the life cycle. Each stage consists of a unique developmental task that confronts individuals with a crisis that must be faced. For Erikson, this crisis is not a catastrophe but a turning point of increased vulnerability and enhanced potential. The more an individual resolves the crises successfully, the healthier that individual’s development will be.
Individuals experiencing a psychological moratorium in advanced industrial societies are often sent to formally structured institutional settings (schools or the military, for example) that will provide them with a time-out from the tasks of adulthood. While the opportunity for young people to experiment with roles, ideas, beliefs, and life-styles can set them on a life course that is rewarding, such opportunity can also result in individuals becoming confused and lost in the identity moratorium (Cote & Allahar, 1994). Cote and Allahar maintain that those youth who cannot bridge the psychological moratorium and who burden the system by deviating in various ways (through truancy or drug addiction, for example) will be embraced by any number of bureaucracies that are poised to take action.

For minor deviations, there are bureaucracies that provide alternating unemployment benefits and work programs, for example. For major deviations, bureaucracies are in place that not only monitor an individual’s behaviour but also seek to correct it, e.g., psychiatric hospitals and prisons. Erikson contends that at this extreme point of alienation an extremely structured situation is imposed on the individual, and this structure provides a much needed identity moratorium, albeit one that confers a negative identity (Cote and Allahar, 1994). Erikson further warned about the permanent harm that can be done when official labels are placed on individuals, particularly when these individuals might be experimenting with their identity (Erikson, 1968). He suggests that the problem and the cure lie not in reinforcing a negative identity but in nurturing a positive one.

Daniel Offer and his colleagues (1988) documented a stereotypical view of adolescence as highly stressful and disturbed. The self-images of adolescents around the world (in the United States, Australia, Bangladesh, Hungary, Israel, Italy, Japan, Taiwan, Turkey and West Germany) were sampled. A healthy self-image characterized at least 73% of the adolescents studied. They appeared to be moving toward adulthood with self-confidence, optimism about the future, and a healthy integration of previous experiences. While there were some differences among the
adolescents, the study found that for the most part they perceived themselves as able to exercise self-control and to cope with life’s stresses. They valued work and school, expressed confidence about their sexual selves, and positive feelings toward their families – not exactly a “storm and stress” portrayal of adolescence.

While most adolescents experience the transition from childhood to adulthood more positively than is portrayed by many adults and media, it is important to remember that they are not a homogenous group of individuals. Most adolescents negotiate the lengthy path to adult maturity successfully; however, a significant minority does not. Gender, age, as well as ethnic, cultural, socio-economic, and lifestyle differences influence the actual life trajectory of each adolescent. Different portrayals of adolescence will emerge depending on the particular group of adolescents described.

Puberty

No matter how adolescence is defined, a central variable is puberty. Puberty is defined as a rapid change to physical maturation involving hormonal and bodily changes that occur primarily during early adolescence (Brooks-Gunn, 1991). These biological changes make the transition from childhood to adolescence developmentally unique: in no other period of life except infancy do so many biological changes occur in such a short a period of time (Montemayor & Flannery, 1990). Montemayor suggests that many of the differences between children and adolescents emerge gradually rather than suddenly. He claims that in most cases, both physiological and psychological developmental change begins in childhood and continues throughout adolescence. Recent studies indicate that pubertal change begins much earlier than previously thought and is a long-term process (Brooks-Gunn & Warren, 1989; Paikoff & Brooks-Gunn, 1990; Rowe & Rodgers, 1990; Brooks-Gunn, 1991, 1992). The implication of these findings is that many individuals classified as children based on age or the absence of
observable secondary sexual characteristics have, in fact, already begun the transition into adolescence.

Puberty is not a single, sudden event (Brooks-Gunn, 1991, 1992). We know when a young boy or girl is going through puberty, but pinpointing its beginning and its end is difficult. Except for menarche, which occurs rather late in puberty, no single marker heralds puberty. For boys, the first whisker or first wet dream is an event that could mark its appearance, but both may go unnoticed.

Montemayor also points out that while there is evidence to suggest that pubertal change influences a variety of adolescent behaviour (i.e., relations with parents, aggression, and sexuality), there are also indications that pubertal effects, by themselves, account for a relatively small proportion of the variance in early adolescent behaviour. Studies indicate that “raging” hormones do not characterize puberty, and that the turmoil once associated with puberty was exaggerated (Brooks-Gunn & Reiter, 1990, Petersen, 1985).

Two conclusions emerge from the recent research on puberty cited above: first, the onset of hormonal pubertal change occurs during middle childhood; and, second, puberty has some affect, but not a powerful affect on adolescent behaviour. These conclusions indicate that puberty alone, and especially observable change, is not an adequate marker of the transition from childhood to adolescence.

Sociological Perspective

It is not a simple task to determine if adolescence is distinctively different from childhood, or if the onset of adolescence is a transitional period. A sociological perspective might possibly help in establishing a clearer understanding of adolescence. Sociologists tend to view adolescence as a product of social expectations associated with a given culture (Cote & Allahar, 1994). They would argue that adolescence and youth are institutions imposed on those
coming of age by forces beyond their control. The difference in the perspective lies in how this imposition is explained. Three theories demonstrate these different perspectives: the functionalist, the subcultural, and the postmodern.

The functionalist views youth and adolescence as a product of social change associated with industrialization. The subcultural perspective argues that because adolescents are not allowed to participate fully in adult culture they develop their own culture in an attempt to find a more meaningful identity. The postmodern view is based on the premise that we have moved out of the modern era, in which science, linear logic, and order prevailed, into a postmodern era that rejects such things. There is no agreed upon postmodern view of youth; it is an emerging perspective. One example of the postmodern view is the argument of Tinning and Fitzclarence (1992), who claim that youth culture is emerging because of the impact of modern information technologies spread by global capitalism and the resultant sense of alienation among youth.

All the perspectives that have been discussed – biological, historical, psychological, and sociological – have depicted adolescents primarily as objects of a society controlled by adults. Adolescents have limited legal rights and economic resources. They are not considered to be fully responsible for their actions, and are generally treated as if they are not capable of acting intentionally – with agency- to find meaning in their lives until they reach adulthood. I believe a shift needs to occur in our understanding of adolescence. It is my contention that adolescents do not merely react to their environment but also influence it. During the transition from childhood to adolescence a wide array of advanced social and cognitive skills emerge that make it possible for adolescents to analyze and to understand themselves, other people, and the world. These new abilities allow adolescents to exert control over their lives to a degree that was not possible only a few years earlier (Caputo, 1995). Two cognitive activities that are especially important are decision-making and critical thinking.
Decision-making

Adolescence is a time of increased decision making – which friends to choose, whether to go to college, who to date, whether to have sex, buy a car, and so on. How competent are adolescents at making decisions? Research tells us that older adolescents (age fourteen to nineteen) often make better decisions than do younger adolescents, who, in turn, make better decisions than children (Keating, 1990). Mann (1997) found that young adolescents are likely to generate options, to examine a situation from a variety of perspectives, to anticipate the consequences of decisions, and to consider the credibility of sources. While older adolescents have increased decision-making skills, these skills are far from perfect, as are those of adults. Indeed, some researchers have found that adolescents and adults do not differ in their decision-making skills (Beyth-Marom et al, 1995).

Such findings have important implications for reducing risk-taking in adolescence and promoting health. For adolescents at risk for Huntington’s Disease who are considering predictive testing, the implications of research into adolescent decision-making may affect how the medical community makes predictive testing available to adolescents who are under the age of eighteen. Unsubstantiated claims about the incompetence of adolescents can lead to paternalism, whereby adults protect adolescents from the consequences of their fallible judgement. In the paternalistic view, adolescents should be denied the right to govern their actions. Adolescents are viewed as not having the capacity to make rational, independent decisions and, therefore, are not entitled to the right of autonomy or free choice.

The testing of adolescents for hereditary late-onset diseases has been judged to be ethically not acceptable in guidelines and directives published by several medical organizations (Elger & Harding, 2000). A number of researchers have taken the position that in special cases minors should be allowed to engage in predictive testing (Elger, 1998; Michie, 1996; Wertz et
al., 1994; Sharpe, 1993). Wertz (1994) recommends that adolescents should be the primary decision-makers when engaged in making reproductive decisions. Sharpe (1993) questions the right to withhold information from minors, and suggests that genetic testing of adolescents needs to be determined on a case-by-case basis. Michie (1996) contends that the arguments against the testing of adolescents are based on the fear of consequences when there is no empirical evidence to substantiate those fears. Elger and Harding (2000) cast concern around adolescent rights to decide about predictive testing because little or no distinction has been made between children and adolescents. They believe a distinction needs to be made between children and adolescents at the age of fourteen years and older. In their discussion regarding testing adolescents for the hereditary breast cancer gene (BRCA1), they argue for age distinction.

Considering the age group of 14- to 17-year-old adolescents, we argue that the reasons against BRCA1 gene testing are not convincing. An analysis of best interest arguments leads to the conclusion that it is best for adolescents to be allowed to decide autonomously about BRCA1 gene testing (2000:333).

It is not only in the arena of predictive testing that questions about adolescent competence in decision-making arise. Some of the ambiguity in the messages about adult status and maturity that society communicates to adolescents take the form of laws dictating that they cannot drive until they are sixteen, vote until they are eighteen, or drink until age nineteen. Yet in some states in the U.S. and some provinces in Canada, fourteen-year-olds now have the legal right to choose the parent with whom they want to live after a parental divorce, and to override parental wishes with regard to such matters as abortion, life-saving medical procedures, and psychiatric care (Volpe et al., 1997). Further, in British Columbia, legal decision-making for health care and reproductive issues falls under Section 17 of the Infants Act, R.S.B.C. 1996, c.223, which provides no specified age of consent for the purposes of obtaining health care. It is important to emphasize that in the absence of parental consent the onus is on the physician to ensure that sufficient explanation is provided, and that the child is capable of giving informed consent. This
is a very heavy burden, and there are few criteria or guidelines to follow. In addition, the issue is somewhat circular in that a doctor-patient relationship must be initiated prior to a discussion about consent.

The ability to make competent decisions does not guarantee that they will be made in everyday life, where the breadth of experience often comes into play (Jacobs & Potenza, 1990; Keating, 1990). For example, driver-training courses improve adolescents' cognitive and motor skills to levels equal to, and sometimes superior to, those of adults. However, driver training has not been effective in reducing the high rate of traffic accidents involving adolescents (Potvin, Champagne & Laberge-Nadeau, 1988).

Perhaps faulty decision-making should not always be blamed for adolescent difficulties; the problem may rest instead in society's orientation toward adolescents. Society may provide many choices, but it fails to provide adolescents with adequate choices. For example, a mathematically precocious ninth-grade girl might abandon mathematics, not because of poor decision-making skills but because of a stronger motivation to maintain positive peer relations that could be threatened if she stayed on the math track. Her decision may not be the consequence of a failure to consider all the relevant information, but rather the result of quite sophisticated thinking about risk-benefit ratios in difficult circumstances that offer limited options. Additionally, adolescents may need more opportunities to practice and discuss realistic decision-making. Liprie's (1993) study of more than 900 adolescents revealed that adolescents were more likely to participate in family decision-making when they perceived themselves as in control of what happens to them, and if they thought that their input would have some bearing on the outcome of the decision-making process.
Critical Thinking

Critical thinking is closely related to making competent decisions (Stanovich, 1993). Although today’s definitions of critical thinking vary, they have in common the notion of grasping the deeper meaning of problems, of keeping an open mind about different approaches and perspectives, and of deciding for oneself what to believe or do (Ennis, 1991; Perkins, Jay & Tishman, 1993; Stanovich, 1993).

Adolescence is an important transitional period in the development of critical thinking (Keating, 1990). The cognitive changes that allow for improved critical thinking in adolescence include increased speed, automaticity, and capacity of information processing, which frees cognitive resources for other purposes; more breadth of content knowledge in a variety of domains; increased ability to construct new combinations of knowledge; and a greater range and more spontaneous use of strategies or procedures for applying or obtaining knowledge, such as planning, considering alternatives, and cognitive monitoring.

Adolescence is an important period in the development of critical-thinking skills. If a solid foundation of fundamental skills (e.g., literacy and math skills) is not developed during childhood, potential gains in adolescent thinking are not likely.

Self-Understanding and Identity

The self is a concept that gradually develops over the human life span. It represents a complex interaction between developing cognitive capacities and socialization experiences; thus, the level of a child’s cognitive abilities coupled with the child’s relationship with parents, peers, and others allows for the development of a portrait of the self. Self-representation shifts from concrete descriptions of one’s psychological and social exterior to more abstract descriptions of one’s psychological interior (Damon & Hart, 1988; Harter, 1986, 1990; Rosenberg, 1979, 1986).
These changes in the nature of self-description are related to the development of cognitive abilities across childhood and adolescence. Piaget's (1967, 1970) stages provide a framework for conceptualizing these changes.

Piaget's Cognitive Theory

Piaget (1954) stressed that adolescents actively construct their own cognitive worlds. Information is not just poured into their minds from the environment; rather adolescents adapt their thinking to include new ideas because additional information furthers understanding. Piaget (1954) also believed that we go through four stages in understanding the world. Each of the stages is age-related and consists of distinct ways of thinking. It is the different way of understanding the world that makes one stage more advanced than another. Knowing more information does not make a child's thinking more advanced in Piaget's view; Piaget believed that a child's cognition is qualitatively different at each stage.

The first stage in Piaget's theory is the sensorimotor stage, which lasts from birth to about two years of age. This is a period during which infants construct an understanding of the world through coordination of sensory experiences (seeing and hearing) with physical motor actions (hence sensorimotor). The second or preoperational stage lasts from approximately two to seven years of age. Children begin to represent the world with words, images, and drawings. During the preoperational period, the young child is only capable of describing observable behaviours or characteristics—giving specific examples rather than generalizations about the self. The young child describes behavioural skills (I can run fast), physical characteristics (I have blond hair), preferences (I love hot dogs), possessions (I have a black dog), and membership categories (I am a girl).

The concrete operational stage runs from approximately seven to eleven years of age. Children can perform operations, and logical reasoning replaces intuitive thought as long as
reasoning can be applied to specific or concrete examples. For instance, concrete operational thinkers cannot imagine the steps necessary to complete an algebraic equation; it is too abstract a concept for this stage of development. What is salient for the nature of self-representation at this stage is a shift from attributes that are observable to those that are more conceptual or trait-like. Trait labels (e.g., smart, happy, popular) represent the newfound ability to classify specific attributes into categories, to form higher-order generalizations about the self. Consequently, specific behaviours such as one's mastery of a number of school subjects may lead one to conclude that one is smart. Trait labels represent a conceptual advance over the previous period because of the capacity to organize observable, behavioural attributes into cognitive concepts about the self.

The formal operational stage, which appears between the ages of eleven and fifteen, is Piaget's fourth and final stage. In this stage, individuals move beyond the world of actual, concrete experiences and think in abstract and more logical terms. As part of thinking more abstractly, adolescents develop images of ideal circumstances. They may think about what an ideal parent is like, and compare their parents with this ideal standard. They begin to entertain possibilities for the future and are fascinated with what they can be. In solving problems, formal operational thinkers are more systematic, developing hypotheses about why something is happening the way it is, and then testing these hypotheses in a deductive fashion.

Piaget's theory emphasizes universal and consistent patterns of formal operational thought. His theory does not, however, adequately account for the unique, individual differences that characterize the cognitive development of adolescents (Overton & Byrnes, 1991). These individual variations in the cognitive development of adolescents have been documented in a number of investigations (Bart, 1971; Neimark, 1982; Kaufman & Flaitz, 1987).

Byrnes (1988a, 1988b) argues that Piaget's theory of formal operational thought can be better understood by considering the distinction between "knowing that" and "knowing how."
“Knowing that” has been called conceptual knowledge or declarative knowledge (Hiebert & LeFevre, 1987; Mandler, 1983). It consists of networks of the core concepts in a given domain, such as biology or physics. “Knowing how” is simply a representation of the steps an individual should follow to solve a problem. It has been referred to as procedural knowledge (Anderson, 1990). For example, in the domain of physics, “knowing that” would consist of understanding the relation between the core concepts of “force” and “mass.” In contrast, “knowing how” would consist of understanding how to solve introductory physics problems using formulas.

Regardless of the debate around Piaget’s theory of formal operations and the nature of adolescent cognitive development, Piaget provides a framework in which to begin to formulate and understand adolescent cognition. Piaget stressed that adolescents are motivated to understand their world because the process of understanding is biologically adaptive. Adolescents actively construct their own cognitive worlds, and to make sense of their world they organize their experience. They can separate important ideas from less important ones and connect one idea to another. They not only organize their observations and experiences, they adapt their thinking to include new ideas because additional information furthers understanding.

Harter’s Ten Dimensions of Self-understanding

With adolescence comes the emergence of descriptions of one’s psychological interior that represent abstractions about the self in the form of beliefs, wishes, emotions, and motives. For example, the adolescent may describe the self as sensitive, moody, obnoxious, and tolerant. Self-representations in the form of abstractions represent a cognitive advance over the previous stage, where the preadolescent could only combine particular behaviours into trait labels (Harter, 1990a). With the advent of formal operational thought, adolescents can now integrate trait labels (e.g., cheerful and depressed) into higher-order abstractions about the self, namely “moody.” To consider oneself “sensitive” one must combine such traits as friendly, understanding, and caring.
Harter (1990a) believes that these unobservable abstract self-attributes represent hypothetical constructs about the adolescent self and, therefore, typically require more inference about one's latent characteristics than do the self-descriptions of children.

The development of self-understanding in adolescence is complex and involves a number of aspects of the self. Susan Harter (1990a, 1990b) provides insightful analysis of adolescent self-understanding. She defines self-understanding as the adolescent’s cognitive representation of the self – the substance and content of the adolescent’s self-conceptions. An adolescent’s self-understanding is based, in part, on the various roles and membership categories that define who adolescents are. Though not the whole of personal identity, self-understanding provides identity’s rational underpinnings (Damon & Hart, 1988).

Harter delineates self-understanding into ten dimensions: abstract and idealistic; differentiated; fluctuating; contradictions within the self; real and ideal, true and false selves (including possible self); social comparison; self-conscious; self-protective; unconscious; and self-integration. As in Piaget’s formal operational stage, many adolescents begin to think in more abstract and idealistic ways. The first dimension of self-understanding – abstract and idealistic – reflects this new capacity. When asked to describe themselves, adolescents will be more likely than children to use abstract and idealistic labels. An adolescent could use an abstract self-definition: “I am a human being.” (“I am indecisive.” “I don’t know who I am.”) An adolescent could also construct an idealistic description of self: “I am a naturally sensitive person who really cares about people’s feelings.” Harter found that not all adolescents describe themselves in idealistic ways, but most adolescents distinguish between the real self and the ideal self (1990a, 1990b).

The second dimension is differentiated self. Adolescent self-understanding becomes increasingly differentiated. Adolescents are more likely than children to describe the self with
contextual or situational variations. Adolescents understand that one possesses different selves, depending on one’s role or the particular context.

The third dimension is contradictions within the self. Once adolescents begin to differentiate the self into multiple roles in different relational contexts, there is the potential for contradictions between these differentiated selves. Adolescents develop the cognitive ability to detect these inconsistencies in the self as they strive to construct a general theory of the self or of their personality (Damon, 1991; Harter & Monsour, 1992).

The fourth dimension is the fluctuating self. Given the contradictory nature of the self in adolescence, it is not surprising that the self fluctuates across situations and across time (Harter, 1990a). Rosenberg (1986) describes the fluctuating nature of the adolescent’s self in terms of the “barometric self.” The adolescent’s self continues to be characterized by instability until the adolescent constructs a more unified theory of self, usually not until late adolescence or even early adulthood.

The fifth dimension includes the real and ideal, true and false selves. The adolescent’s emerging ability to construct an ideal self can be perplexing for the adolescent. While the capacity to recognize a discrepancy between real and ideal self represents a cognitive advance, too great a discrepancy can, as humanist theorist Carl Rogers (1950) believed, indicate maladjustment.

Researchers have found that the discrepancy between the real self and ideal self is greater in middle adolescence than in early or late adolescence (Strachen & Jones, 1982). While Rogers (1950) considers a strong discrepancy between the ideal and real selves as maladaptive, others argue that this is not always true, especially in adolescence. Markus and Nurius (1986) found that an important aspect of the ideal or imagined self is the possible self. The possible self is what individuals might become, what they would like to become, and what they are afraid of becoming. Thus, the adolescents’ possible selves include both what adolescents hope to be as
well as what they dread they will become. In this view, the presence of hoped-for as well as
dreaded selves is psychologically healthy, providing a balance between positive, expected selves
and negative, feared selves. The attributes of future positive selves (getting into a good college,
being admired, having a successful career) can direct future positive states, while attributes of
future negative selves (being unemployed, being lonely, not getting into college) can identify
what is to be avoided in the future.

Harter and Lee (1989) found that adolescents could distinguish between their true and
false selves. Adolescents are most likely to show their false self in romantic or dating situations
and with classmates; they are least likely to show their false self with close friends. Adolescents
display a false self to impress others, to try out new behaviours or roles, and because others do
not understand their true self and thus force them to behave in false ways (Harter, 1990b).

The sixth dimension of self-understanding is social comparison. Some developmentalists
believe that adolescents are more likely than children to use social comparison to evaluate
themselves (Ruble et. al., 1980). However, adolescents’ willingness to admit that they engage in
social comparison to evaluate themselves declines in adolescence because they view social
comparison as socially undesirable. They think that acknowledging their social comparison
motives will endanger their popularity (Harter, 1990a).

The seventh dimension of self-understanding is self-consciousness. Adolescents are more
likely than children to be self-conscious about and preoccupied with their self-understanding. As
part of their self-conscious and preoccupied self-explorations, adolescents become more
introspective. However, the introspection is not always done in social isolation; sometimes
adolescents turn to their friends for support and self-clarification, obtaining their friends’
opinions of an emerging self-definition. Rosenberg (1979) found that adolescents’ friends are
often the main source of reflected self-appraisals, becoming the social mirror into which
adolescents anxiously stare.
The eighth dimension of self-understanding is self-protectiveness. Adolescents' self-understanding includes more mechanisms to protect the self (Harter, 1990a; 1990b). Although adolescents often display a sense of confusion and conflict stimulated by introspective efforts to understand the self, they also call on mechanisms to protect and enhance the self. In protecting the self, adolescents are prone to denying their negative characteristics. For example, in Harter's investigation of self-understanding, positive self-descriptions (attractive, fun-loving, sensitive, affectionate, inquisitive) were more likely to be ascribed to the core of the self, indicating more importance, whereas negative self-descriptions (ugly, mediocre, depressed, selfish, nervous) were more likely to be ascribed to the periphery of the self, indicating less importance (Harter, 1986). Adolescents' tendency to protect themselves fits with the earlier description of adolescents' tendency to describe themselves in idealistic ways.

The ninth dimension of self-understanding is the unconscious. Adolescents' self-understanding involves greater recognition that the self includes unconscious as well as conscious components, a recognition not likely to occur until late adolescence (Selman, 1980). That is, older adolescents are more likely than younger adolescents to believe that certain aspects of their mental experience are beyond their awareness or control.

The last dimension of self-understanding is self-integration. Adolescents' self-understanding becomes more integrative, with the disparate parts of the self more systematically pieced together, especially in late adolescence. Older adolescents are more likely to detect inconsistencies in their earlier self-descriptions as they attempt to construct a general theory of self, an integrated sense of identity (Harter, 1990b; Selman, 1980).

Because the adolescent creates multiple self-concepts in adolescence, the task of integrating these varying self-conceptions becomes problematic. At the same time that adolescents are faced with pressures to differentiate the self into multiple roles, the emergence of formal operational thought presses for integrations and the development of a consistent, coherent
theory of self (Harter, 1990b). These budding formal operational skills initially present a liability because they first allow adolescents to detect inconsistencies in the self across varying roles, only later providing the cognitive capacity to integrate such apparent contradictions. Researchers have found that fourteen- to fifteen-year-olds not only detect inconsistencies across their various roles (with parents, friends, and romantic partners, for example) but are also much more troubled by these contradictions that younger (eleven- to twelve-year-olds) and older (seventeen- to eighteen-year-olds) adolescents (Damon & Hart, 1988; Harter, 1986).

Erikson’s Theory of Adolescent Development

An important concept related to the self is that of identity. Erikson’s (1968) theory of adolescent development outlines eight stages of development that unfold as we go through the life cycle. Each stage consists of a unique developmental task that confronts individuals with a crisis that must be faced. For Erikson, this crisis is not a catastrophe but a turning point of increased vulnerability and enhanced potential. The more an individual resolves crises successfully, the healthier that individual’s development will be.

Trust versus mistrust is Erikson’s first psychosocial stage, which is experienced in the first year of life. A sense of trust requires a feeling of physical comfort and a minimal amount of fear and apprehension about the future. Trust in infancy sets the stage for a lifelong expectation that the world will be a good and pleasant place to live.

Autonomy versus shame and doubt is Erikson’s second stage of development, occurring between the ages of one and three. After gaining trust in their caregivers, infants begin to discover that their behaviour is their own. They start to assert their sense of independence or autonomy. They realize their will. If infants are restrained too much or punished too harshly, they are likely to develop a sense of shame and doubt.
Initiative versus guilt is Erikson’s third stage of development, occurring during the preschool years. As preschool children encounter a widening social world, they are increasingly challenged. Active, purposeful behaviour is needed to cope with these challenges. Children are asked to assume responsibility for their bodies, their behaviour, their toys, their pets. Developing a sense of responsibility increases initiative. Uncomfortable guilt feelings may arise, though, in children who are irresponsible and are made to feel anxious. Erikson has a positive outlook on this though: He believes that most guilt is quickly compensated for by a sense of accomplishment.

Industry versus inferiority is Erikson’s fourth developmental stage, occurring approximately in the elementary school years. Children’s initiative brings them in contact with a wealth of new experiences. As they move into middle and late childhood they direct their energy toward mastering knowledge and intellectual skills. At no other time is the child more enthusiastic about learning than at the end of early childhood’s period of expansive imagination. The danger in the elementary school years is the development of a sense of inferiority – of feeling incompetent and unproductive.

Identity versus identity confusion is Erikson’s fifth developmental stage, which individuals experience between 12 years-of-age and 20 years-of-age. At this time individuals are faced with finding out who they are, what they are all about, and where they are going in life. Adolescents are confronted with many new roles and adult statuses – vocational and romantic, for example. If the adolescent explores such roles in a healthy manner and arrives at a positive path to follow in life, a positive identity will likely be achieved. If an identity is pushed on the adolescent by parents, or if the adolescent does not adequately explore many roles and a positive future path is not defined, then identity is challenged.

Intimacy versus isolation is Erikson’s sixth developmental stage, which occurs during the early adulthood years. At this time individuals face the developmental task of forming intimate
relationships with others. Erikson describes intimacy as finding oneself yet losing oneself in another. If the young adult forms healthy friendships and an intimate, close relationship with another individual, intimacy will be achieved; if not, isolation will result.

Generativity versus stagnation is Erikson’s seventh developmental stage, and is experienced during middle adulthood. By generativity Erickson means assisting the younger generation in developing and leading useful lives. Stagnation refers to the feeling of having done nothing to help the next generation.

Integrity versus despair is Erikson’s eighth and final developmental stage, which is experienced during late adulthood. In our later years, we look back and evaluate what we have done with our lives. Through many different routes the older person may have developed a positive outlook in most or all of the previous developmental stages. If this is the case, the retrospective glances reveal a life well spent, and the person feels a sense of satisfaction – integrity is achieved. If the older adult resolved many of the earlier developmental stages negatively, the retrospective glance will likely engender doubt or gloom – the despair Erikson talks about.

Two core ingredients in Erikson’s theory of identity development are personality and role experimentation. As indicated earlier, Erikson believes that adolescents face an overwhelming number of choices, and at some point during youth enter a period of psychological moratorium. During this moratorium they try out different roles and personalities before they reach a stable sense of self.

As adolescents come to realize that they will be responsible for themselves and their own lives, they search for what those lives are going to be. Many parents and adults, accustomed to having children go along with what they say, may be bewildered or incensed by the rebelliousness and the rapid mood changes that can accompany adolescence. Erikson believes
that adults need to give adolescents time and opportunity to explore different roles and personalities. He believes that in turn most adolescents will eventually discard undesirable roles.

The development of an integrated sense of identity is a long, complex, and difficult task. In Western culture, adolescents are expected to master many different roles. Most adolescents have some difficulty handling all these roles competently. Bourne (1978) explicates several dimensions of Erikson's theory of identity, stressing that identity is the adolescent's adaptation to the society in which she/he lives. Bourne emphasizes the dynamic nature of Erikson's theory: Erikson believes that identity formation arises from childhood identifications with adults but absorbed in new configurations that are, in turn, dependent on society's roles for youth. Further, Erikson emphasizes the mutual relationship of adolescents with their social world and community. Identity development is not just an intrapsychic self-representation but involves relationships with people, community, and society. Bourne (1978) also ascribes an existential status to Erikson's theory: Erikson believes that adolescents seek meaning in their own lives as well as the meaning of life in general.

Contemporary Thoughts on Identity

Contemporary views on identity development offer several important conclusions. First, identity development is a lengthy process, and in many instances is a more gradual, less cataclysmic transition than Erikson's "crisis" implies (Baumeister, 1991). Second, identity development is extraordinarily complex (Marcia, 1987; 1989). Identity formation neither begins nor ends in adolescence. It begins with the appearance of attachment, and continues with the development of a sense of self and the emergence of independence in infancy, reaching its final phase with a life review and integration in old age. What is important about identity development in adolescence, especially late adolescence, is that, for the first time, physical development, cognitive development, and social development advance to the point at which the individual can
sort through and synthesize childhood identities and identifications to construct a viable path toward adult maturity. However, resolution of the identity issue during adolescence does not mean that identity will be stable throughout the remainder of life.

Identity formation does not happen neatly, nor is it generally a cataclysmic event. Identity development gets done in bits and pieces. Decisions are not made once and for all but have to be made again and again. The decisions may seem trivial at the time: whom to date, whether or not to break up, whether or not to have intercourse, whether or not to take drugs, whether or not to go to college or finish high school or get a job, and so on. Over the years of adolescence, the decisions begin to form a core of what the individual is all about as a human being – what is called his or her identity.

Adolescents also begin to develop more abstract self-characterization, and self-concepts become more differentiated and better organized. They begin to view themselves in terms of personal beliefs and standards, and less in terms of social comparisons (Harter, 1998). We also know that adolescents evaluate themselves both globally and along several distinct dimensions – academics, athletics, appearance, social relations, and moral conduct (Marsten & Hubbard, 1995) – and that the link between specific dimensions of self-concept and global self-worth vary across domains. For example, appearance is most important for overall self-esteem, especially among females (Usmani & Daniluk, 1997). There is also evidence that adolescents’ self-conceptions differ across contexts, and that teenagers see themselves differently when they are with peers than when they are with parents or teachers (Harter, 1998). In general, global self-esteem is stable during adolescence. Harter reports that global self-esteem may increase slightly over the period of adolescence (1998), becoming stable with age (Alaker & Olweus, 1992; Harter, 1998). Research also indicates that some adolescents show high levels of stability in self-esteem while others do not (Diehl et al., 1997), and that self-esteem varies according to ethnicity and
gender. Across all groups, however, high self-esteem is related to parental approval, peer support, adjustment, and success in school (DuBois et al., 1998, Luster & McAdoo, 1995).

Cultural Agency

One of the important ways in which adolescents contribute to their own development is by choosing the environments and social contexts in which they participate. They make choices about friends, what subjects to take in school, how to spend leisure time, and in a multitude of other areas, all of which affect future developmental pathways. Adolescents are involved in the production and management of culture in youth peer groups (Wulff, 1995). Early anthropological studies viewed youth as on their way to adulthood – in the process of learning for later challenges – rather than producing their own culture that might not last in the long run but could still be significant for them at the time (Wulff, 1995). James (1993) states that culture, portrayed in terms of a unified system of meaning, privileges the voices of the powerful and excludes the voices of women, adolescents, and children. Wulff contends that the concept of culture needs to be reconceptualized in a way that includes the issue of voice and agency in the context of children’s lives.

Hannerz (1992) argues that people negotiate culture, or cultural processes, and are formed by them to a certain extent. Wulff (1995) suggests that where young people form these cultural processes a youth culture emerges. This idea of youth as cultural agent is grounded in the idea of agency cited by Giddens, and is defined as “the stream of actual or contemplated causal interventions of corporeal beings in the ongoing process of events-in-the-world (1976:75).” Wulff links the concept of agency to that of practice.

James (1993) examines the notion that children may have a different kind of knowledge, that they may be preoccupied with things that adults are unaware of. James describes how she had to “learn to talk again” in a virtually new vocabulary in order to make conversation with the
children she was studying. She found that children challenged the limits of social action established by adults through restyling language (especially terminology, rhythm, and cadence) thereby signifying their identities as “child” and/or “youth.” According to James, it is through developing their performative language skills that children progress into youth culture, which continues to be united with children’s culture by the experience of marginality.

As these anthropological studies demonstrate, adolescents are not viewed as “on their way to adulthood” but as producing their own culture (Wulff, 1995). How might the recognition that adolescents navigate their own culture affect the study of youth at risk for HD? Additionally, if the voice and agency of the adolescent is marginalized by social processes (James, 1993), and in effect absent in social processes, will this affect our approach to an understanding of adolescents at risk for HD?

**Youth Culture**

Youth culture in North America is often associated with teenagers – people between the ages of thirteen and nineteen. Young teenagers are most likely attending school; older teenagers have probably completed school. The experience of the younger and the older teenager will be quite different. Klien (1990) contends that we need to re-evaluate current conceptualizations of life-stage theory to include a stage of youth. Like all other life stages, youth has certain characteristics that distinguish it from the life phases of adolescence and young adulthood. He claims adolescence is characterized by the individual’s attempt to develop a world outlook that will carry the adolescent through life and guide the adolescent’s expectations. Young adulthood is a time when the adolescent tasks of development are resolved. Young adults are expected to be gainfully employed, financially independent, to have a direction in life, and to be working toward their life goals.
In the midst of late twentieth century politico-economic transformations, new youth cultural forms emerged (and continue to emerge) in North America. Attention must be devoted to the analysis of new forms of youth cultural production; inherent in the concept of youth culture is the recognition of the capacity for cultural agency. Goodenough (1994) argues that culture is more usefully located in activities rather than communities – in the expectation people have of interaction and the standards of evaluation operating within a particular situation. He states that even in the most isolated of communities, people are engaged in a variety of activities with differing organizational requirements; people must therefore engage different sets of situated understandings and expectations. Amit-Talai (1995) believes that people have to be multi-cultural.

Youth cultural production occurs at home, at school, at work, at play, on the street, with friends, teachers, parents, siblings and bosses, draws elements from home-grown as well as transnational influences, and intertwines with class, gender, ethnicity and locality with all the cultural diversity that such a multiplicity of circumstances compels. (1995:166)

One aspect of cultural agency that is not adequately addressed by the research is the role of the medical community (Hamburg, 1995). Do adolescents have agency within the biomedical community? Do they have a voice? In what way has the biomedical community applied the “official labels” that Erikson (1957) warned as conferring a “negative identity” on adolescents? In what way does the medical community confer a negative identity on the adolescent at risk for HD?

Psychopathology and Adolescence

For much of the twentieth century in the United States and other Western cultures, adolescence has been perceived as a problematic period in the human life span that youth, families, and society had to endure. What are the problems of youth? Hill and Fortenberry (1992) note that adolescents are described as major players in the multiple “epidemics” that beset the
country: pregnancy, sexually transmitted diseases, drug abuse, and suicide are a few of the more obvious examples.

Fabrega Jr. and Miller (1995) believe that this picture of contemporary adolescence is partly substantive and partly constructionist, and refers to real changes in society and to symbols that elaborate on and explicate the consequences of the changes. They believe that there are four related ideas implicit in this picture of adolescence and its psychopathology. First, is the idea of a new structural formation: a social group (adolescents) produced as a by-product of social differentiation and complexity linked to industrial capitalism. Second, is the idea of self-consciousness: adolescents become more aware of their unique position in society and of themselves as a group. Third, is the idea that adolescents are exploited and manipulated by capitalists who see a new teenage market for leisure. Fourth, is the idea of pathology linked to adolescence as a social stage, a theme presented in the writings of Hall and still prevalent in much of the literature on adolescence. In their review of adolescent development, Steinberg and Morris (2001) conclude that the scientific study of adolescent development has always had as part of its implicit and explicit agenda the goal of describing, explaining, predicting, and ameliorating problematic behaviour. He argues that this view prevails despite evidence that the majority of teenagers weather the challenges of the period without developing significant social, emotional, or behavioural difficulties (Stienberg, 1999).

The Medicalization of Adolescence

What is “medicalization? Conrad (1992) refers to medicalization as the process whereby a condition comes to be defined and understood as an illness and is therefore moved into the sphere of control by the medical profession. Medicalization occurs at three levels: (1) the conceptual, which involves the use of medical language and models to understand a condition or process; (2) the interactional, in which, as part of doctor-patient interaction, a physician defines a
condition as medical or treats a social problem as though medical in nature; and (3) the institutional, in which the condition or process comes under the jurisdicentral control of the medical profession (Conrad, 1992; Conrad & Schneider, 1992; Zola, 1972). Hill and Fortenberry (1992) argue that there is a tendency in American society to seek biological explanations for a wide range of psychosocial phenomena. They use examples of symptom complexes like chronic fatigue syndrome and hypoglycaemia as examples of apparently common syndromes for which there is little data to support the biological explanations that have been proposed.

Locke (1986) examined the way in which medicalization in Japan is shaped by an ideology in which contemporary biomedical ideas appear to be modified by traditional values. For instance, individuals or families are usually held responsible for problems that have a broad range of etiological components – sickness of the elderly, behavioural problems in children, and depression in housewives, for example. Locke explores how the structure of the medical system in Japan also influences the process of medicalization. She provides three case histories of “school refusal syndrome” and gives various popular interpretations for the occurrence and treatment of this syndrome. She concludes that medical anthropology needs to pay critical attention to “macro-structural questions, the role of power in social life, and the way in which biomedicine is culturally constructed” (1986:107).

In Japan it is believed that individual needs are best kept contained and controlled for the sake of group co-operation, advancement, and social harmony. Locke states that treating refusal to go to school as a syndrome – a familial-somatic problem – serves to reinforce the dominant ideology of the society. She hypothesizes that children are making a request through their refusal. The request may eventually become distorted or expressed as serious illness, but initially

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3 School refusal syndrome refers to a type of behaviour that is classified by the Ministry of Education in Japan. Children from primary school through to college act as though they wish to go to school, but in fact refuse to go, and remain in their house and often in their bed all day. The problem is treated as a familial-somatic one. Lock (1986) states that although social aspects of the problem are acknowledged, individuals and their families are held morally responsible for “failing to adjust adequately to the larger social system” (p. 107).
they are asking to be recognized for what they are and not for what they represent to their father, mother, schoolmates, teacher, or larger society.

Locke, like Fabrega Jr. and Miller (1995), concludes that the form that medicalization takes is culturally shaped. She contends that in order for us to develop sophisticated theories using a critical perspective we need to acknowledge that capitalist states, professions, and bureaucracies are composed of real people, and that while many of these people act blindly, not all of them do.

Whether medical professionals act blindly or not, Offer and Howard (1981) and Hartlage et al. (1985) found that psychiatrists, psychologists, and other professionals working with teenagers described normal or mentally healthy adolescents as moody and rebellious, and in constant emotional turmoil. They concluded that normal teenage behaviour bordered on delinquent, and that teenagers had poor coping abilities. In the *Textbook of Psychiatry* (1989) under the heading of “Adolescence Proper,” a number of significant factors of adolescent turmoil are labelled as developmentally deviant and at high risk for psychopathology in adult years.

Hill and Fortenberry (1990) conducted two surveys to identify the features of images of adolescents. The first survey canvassed 103 medical students who were asked, “When the word ‘teenage’ or ‘adolescent’ is mentioned, what is your first image or impression?” Over half (58.3 percent) of the medical students’ responses fell into the category of negative images. The second survey had a more diverse group of respondents yet each survey demonstrated that adults perceive adolescents in negative ways. The medical students appear to have had the most negative perceptions. To what extent does medicine and the medical profession contribute to the growing public stereotype of adolescence as morbid and pathological?
According to Western biomedicine, psychopathology encompasses specific psychiatric disorders and general problems of adaptation. From a cultural anthropological standpoint, psychopathology can be equated with behaviour that in local systems of meaning is judged as deviant and/or as a breakdown in adaptation. Fabrega Jr. and Miller (1995) suggest that there is a logical discontinuity between the biomedical and local cultural frame of reference. Each facet of what in any society is characterized as psychological is under the sway of culture. Anxiety, depression, and psychosis can be viewed from physiologic and neuropsychologic standpoints as universal, but how they are realized reflects cultural influences.

There are two approaches to the study of how culture shapes psychopathology (Shweder, 1991). The first approach holds that family structure and child rearing practices are the forces that lead to the formation of personalities characterized by distinctive styles of psychological and social function, malfunction, and conflicts. It is this view that characterizes the principles of Western psychology. The second approach, termed “cultural psychology,” examines human psychologies not through the perspective of Western psychology but in terms of the logic and meaning of a particular culture. The cultural psychology approach places less emphasis on the dynamics of how behaviour and “pathology” are produced and more on the construction, meaning, and organization of culturally appropriate behaviour.

Fabrega Jr. and Miller (1995) conclude their review of the development of adolescent psychiatry by considering three problem areas of adolescent behaviour: eating and bodily appearance, psychological identity, and autonomy and social aggression. In the official diagnostic schema of psychiatry, these problems are represented as anorexia nervosa, dissociative disorders, and oppositional and conduct disorders. The contention is that these three conditions are influenced by social and cultural changes brought about by recent historical
developments linked to Anglo-American and, more generally, western industrial capitalism. They conclude that psychiatry generally – and adolescent psychiatry in particular – has played a complex role in the medicalization of adolescent problems associated with modern capitalism.

Adolescence as Culture-Bound Syndrome

Hill and Fortenberry (1992:73) argue that adolescence itself has been medicalized into a condition that is inherently pathological. They explore adolescence as a “culture-bound syndrome” – a “constellation of symptoms, which has been categorized as a disease.” They contend that the development of the concept of the “at-risk” or “high-risk” adolescent has made adolescence a marker for disease, i.e., a symptom. They propose that culture-bound syndromes in highly differentiated societies might be classified taxonomically by means of the distinctive cultural identity system domains of age, gender, family, vocation, and ethnicity. According to this taxonomy, American adolescence is an age-based, culture-bound syndrome.

The Dilemma of Medical Professionals

G. Stanley Hall’s storm and stress model of adolescence has been recast as a symptom complex requiring diagnosis. Hill and Fortenberry (1992) quote Michael Baizerman (1988) who writes,

The medical model of diagnosis, pathogenicity, and therapy used therein has become the lens used almost universally to view youth as behaviour and life as facts and living as troubles, problems, or illness. Being a kid, with its orientation to exploration, adventure, and risk is seen as a “condition” needing control and prevention. (1988:131)

If medical professionals are to perceive themselves as advocates for the well-being of adolescents, it is necessary for them to consider their contribution to the growing public stereotype of adolescents as morbid and pathological. As Fabrega Jr. and Miller (1995) point out,
“In medicalizing adolescent problems the discipline helps to subjugate and control but in casting itself as healers and advocates it seeks to mediate and restore.”

Constructing a New View of Adolescence

It is my contention that adolescents are individuals with inherent needs and the capacity to come of age in their own right. The young can be seen as acting intentionally – with agency – to find meaning in their lives. Growing through adolescence is a non-linear process that involves choosing between alternative courses of action. In order to advocate for adolescents we need to begin to understand how adolescents are active agents engaged in the production and management of meaning in their own lives. It is important for healthcare professionals to explore with adolescents what their experience of adolescence is. It may well be that what is perceived as behavioural disturbances among adolescents – disorders with various sets of symptoms – are actually responses to problems produced by culture.

In addition to views about adolescents, healthcare providers have views about patients. Waxler-Morrison et al. (1990) contend that healthcare professionals and patients can bring different notions to the clinical encounter. Professionals bring the biomedical culture as well as their own ethnic and sociocultural background. Patients bring their own ideas about the biomedical model and their own set of beliefs and values about illness. The authors argue that there are often discrepancies between these two perspectives in the explanation of disease and illness, as well as in the expectations of how each should behave and of treatment results. The issue is not that viewpoints differ but that these differences can lead to misunderstanding. It is important for healthcare professionals to explore with their patients salient issues such as belief about illness, expectations of treatment, and how illness is managed in daily life.

Great Transitions (1995), the Carnegie Council report on adolescence, concludes that institutions have not provided the support that adolescents need. It argues that adolescents are not
receiving the education, direction, and assistance that would help them deal with the stresses and risks inherent in this age group in post-industrial times. Five institutions are identified as critical to young adolescents: the family, schools, youth-serving groups, healthcare organizations, and the media. All are chided for falling behind in their “vital functions,” and the report calls on them to become “strengthened in their respective roles and linked in a mutually reinforcing system of support for adolescents.” It concludes that society has stereotyped adolescents as “difficult and unpleasant,” and that “family, schools, health sector, community organizations, and media have not provided the necessary guidance and protection.”

The idea of adolescence that is constructed by the community and the organizations of the community (i.e. biomedical community) in which the adolescent lives, may be an inaccurate representation of adolescents and may reflect very little of the culture – voice and agency – that our youth make their own. The stereotyping of adolescents by the society at large, the medicalizing of adolescence by the biomedical culture, and the view of adolescence as a problem continues to marginalize youth and lock them in the “storm and stress” model. In light of this review, any particular behaviour must be considered in terms of how it demonstrates a strategy of coping with the cultural and social context rather than being labelled as pathology. Only when we begin to explore the voice and agency of adolescent culture that “the experience of these kids in growing up can be treated more accurately, more sympathetically, and to the extent possible, create some restraint with respect to the more damaging influences” (Hamburg, 1995:7).

Summary

Adolescence is a transitional period in the human life span. It is a segment of continuing human development that is best characterized as an individual process influenced by cognitive, developmental, and the socio-cultural context of the adolescent’s life. As researchers carefully examine adolescents’ lives, they increasingly recognize the complexity of adolescent
development. Because of this complexity, no single developmental model is likely to fit all adolescents. Adolescents must be viewed as a heterogeneous group because different portrayals of adolescence emerge, depending on the particular set of adolescents being described.

The majority of adolescents today successfully negotiate the path from childhood to adulthood. However, too many of today’s adolescents are not provided with adequate opportunities or the support required to make a successful transition to adulthood. In order to advocate for adolescents we need to begin to understand how adolescents are active agents engaged in the production and management of meaning in their own personal and social lives. It is my belief that it is important for healthcare professionals to explore with adolescents what their experience of adolescence is.

Adolescents living in families with HD and therefore at risk for HD represent a group with significant additional stresses. Little information is available concerning the experience of this group of adolescents. Parents as well as medical professionals need to understand the experience of these at-risk adolescents in order to provide support and medical services that will enable them to cope with the challenges of this genetic disease. The research presented in this paper begins to explicate the adolescent experience of living at risk for HD, as well as address how parents and professionals can assist adolescents in that experience.

Summary of Research on Adolescents and Huntington’s Disease

As Hayes (1992:1450) states, “Many professionals have difficulty viewing genetic issues in a family context. Nevertheless the diagnosis of Huntington’s Disease affects every member of the family.” Indeed, the onset of HD often occurs after individuals have married and had children. Consequently, most offspring of HD carriers will live a major part of their lives with the knowledge of being at 50% risk of developing the disease (Jensen et al., 1993). There is little
research on how the presence of HD in a family affects the members of that family, especially the children.

In one of the first studies that addressed the experience of HD within the family, Barette and Marsden (1979) described these families as experiencing “widespread anxiety and suffering.” Their study attempted to establish how and when family members discovered HD in the family, what effect this knowledge had on them, and their reaction to a possible predictive test for the disease. The study focused on the potential impact of predictive testing for those at risk.

The study was based on 153 responses to questionnaires mailed to spouses or “first-degree relatives” of individuals with HD. There was no systematized data on the reason for failure to reply. The researchers state that people did not reply because some found the questions too emotionally disturbing to consider and answer, or the non-factual questions were too difficult. No replies were accepted from people with symptoms of HD. Of the 153 informants, 100 were visited in their homes for additional information. The questions in the questionnaire were phrased in an open-ended manner, asking for subjective opinions about the affected persons feelings as well as their own feelings. For example, one question was, “What effect did discovering that the disease is hereditary have on you or your relative at the time?”

The results of the study showed that most people learned about the disease from hospital consultants, not from their family. The study was done prior to linkage or predictive testing and therefore it was difficult to establish a family history of HD, and problems of misdiagnosis were frequent. Many of the respondents who had been informed of the illness had not been informed of the hereditary nature of the disease and the consequence to the family. Only 66% of the

4 This study was conducted prior to the discovery of a linked marker for the HD gene. The 1983 discovery of a linked marker made it possible for at-risk individuals with informative family structures to know with high likelihood whether or not they would develop the disease.

5 First-degree relatives are the spouses, children, and siblings of HD-affected individuals.
respondents answered the question concerning how they proposed to tell their children about the hereditary nature of the disease. The respondents reported that the question produced “great mental anguish.”

The study identified two aspects of the problems faced by HD families that stood out as warranting further study: first, the means of genetic control of the illness\(^6\); and, secondly, the avenues available and required to help such families. The researchers attempted to create a strong argument against reproduction for people at risk for HD, and to assign the responsibility for control of reproduction to the whole family unit—“affected and unaffected members being equally responsible for containing the procreative attitudes of their sibs and children” (1979:335). The authors of this study did not include affected individuals in their questionnaire, but held them partially responsible for future generations.

The aim would be to apprise those at-risk of the facts, but with the added knowledge that responsibility for control of the illness rests upon all members of that family unit and with the hope that a corporate decision would be made and enforced by the family unit itself. (1979: 335)

The authors also recognized that “the disease with its implications was often too enormous and overwhelming, and there was a continual and repeated need for support” (1979:335). On the one hand, families were to be responsible for the actions of one another, while on the other hand they were in such an overwhelming state that they required continual support. The study contributed to the beginning of the recognition that HD, because of its hereditary nature, is a disease that impacts the whole family.

Tyler et al. (1983) conducted a study in Wales that examined the relationship between disease state and family breakdown and stress. Their goal was to measure the extent of unmet need, and to suggest ways in which the clinical management of HD families could be improved.

\(^6\) Genetic control in this research means regulation of procreation in HD families.
The ninety-two families in this study were gathered through a larger population survey, and the information provided in the study was from hospital records, structured psychiatric interviews, as well as from the "primary care agents" of the HD patients. The study was significant because it was one of the few studies that examined the effects of HD on children in an HD family. Although the information was not obtained from the children directly, the authors concluded that patients and their families seemed to suffer an impoverished quality of life, particularly in their relationships: few children felt close to the affected parent, even when the disease was of late onset. Adult children interviewed (primary care agents in this study) spoke of continuous quarrelling and fights, which caused them to leave the family as soon as possible, often by marrying very young. The authors contend that there is strong evidence that "choreics," in general, do not make good parents.

It is difficult to determine the reliability of the results of this research: twenty of the ninety-two patients studied were dead by the time of the social survey, and fifteen of the patients had been permanently hospitalized. The interviews with the "primary care agents" were primarily responses to close-ended questions, or were ratings of symptoms of stress. The healthy parent, "if strong," was considered an important person to compensate for the adverse effects of the affected parent. They also state that the picture is a variable one; some children may "escape relatively unscathed, others suffer traumatic experiences" (1983:136). As in the Barette and Marsden study (1979), the authors conclude that the patient as well as the family needs a great deal of support. Tyler suggests that medical and social agencies lack an understanding of the problems of these families.

The Tyler study contributed to increasing the understanding of the familial nature of HD and the impact of the disease on the family. The researchers were able to determine that the

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7 Chorea (from the Latin choreus) is a term used to describe involuntary dance-like movements. The association of these movements with the disease first identified by George Huntington led it to become known as Huntington's Chorea.
The amount of unmet need for HD families is high, "partly because of ignorance about entitlement to benefits and services and partly because of lack of facilities" (1983:136). The study suggested a "management program" that contained a "reaching-out" policy combined with improved education and training of healthcare workers.

Power (1984) conducted the first study to explore the emotional reaction and coping of adolescents (ages thirteen to sixteen) to their parent's neurological illness. The affected parents in this study had been diagnosed with either Huntington's Disease or Multiple Sclerosis (MS). The researcher interviewed thirty-seven HD and MS families for ten sessions. The methodology was defined as an "observational method with a particular focus on the attitudes and behaviours of family members" (1984:46). The article described the emotional reaction of the adolescent children to parental disability, and compared the coping styles of these children. In conclusion, intervention approaches were suggested that encouraged the family attributes of understanding, expectations, and acceptance.

This is the first study of Huntington's Disease to acknowledge the family as representing an important social context in which illness occurs and is dealt with. However, the author bases his belief about the "staggering effect" of parental disability upon the adolescent on clinical observations that are undocumented by research. The chronically ill parents were initially seen at the researcher's office for two sessions, and then the entire family was seen together for six home visits and two office visits. The adolescents who are the focus of this study were never interviewed alone. Barker (1990) in his book on clinical interviewing of children and adolescents suggests that adolescents should be interviewed first or in the company of the family. He contends that if their parents or other adults are interviewed first, the adolescents may conclude that unfavourable reports about them have been made before they have had a chance to state their points of view.
Power's analysis suggests that within the families studied the common reaction of all the young people was anxiety, although he found that this emotion was heightened in the adolescents in families with Huntington's Disease. He also found feelings of disappointment, shame, and stigmatization among all the adolescents. Of the thirty-eight young people in the MS families, thirty-one showed no observable change in their basic lifestyle while the majority of young people in the HD families had more intense and extensive reactions of tension and disappointment. Living with the reality of having a 50:50 chance of being like the HD-affected parent was difficult for twenty-two of the thirty-one adolescents from the HD families. According to the author "they seemed very angry over the illness situation" (1984:48). Power did not explain what comprised "the illness situation." Is it the parent's illness, the family's inability to cope, the adolescent's reaction to the parent's inability to cope, the adolescent's sense of similarity to the affected parent, the adolescent's understanding of risk status? The research did not explore the adolescents' feelings about their own risk status. The focus was on how the children coped with a disabled parent, and their feelings concerning the impact of the parent's illness on them.

When discussing coping styles, Power refers to Lazarus' "emotion-focused" coping method. Lazarus categorizes coping mechanisms into two categories: "problem focused" and "emotion focused." With the former an individual tries to change the situation for the better, while the latter would include things one does or says to oneself to feel better, such as minimizing the problem, thinking of something else, and even distancing oneself from the stressful situation. Lazarus believes that well-functioning people use a mixture of problem-focused and emotion-focused coping methods (Lazarus, 1984). Power found that adolescents from both MS and HD families who were coping well used some form of emotion-focused coping as well as "positive denial." Other positive coping mechanisms reported by these adolescents included the opportunity to talk about the parent's illness with the other parent or
with other relatives. The adolescents who reportedly were coping poorly not only used drugs but also displayed “giving up” behaviours in the form of withdrawal from accustomed activities or an “unwarranted dependence” on the well parent.

The author states that the Q-sort results and interview observations suggest that three family behaviours have the strongest influence on how the adolescent copes: family understanding of disability, family expectations for the patient and children, and family acceptance. Family understanding of disability did much to ease patient and family distress precipitated by the uncertainty of the illness; it conveyed to the adolescents that many family activities could continue normally. An awareness of the residual physical and emotional assets of the parent was reported to bring hope to the adolescents and alleviate some of the emotional distress resulting from changes in accustomed activities. If information was not communicated, or the family did not understand the information that was communicated to them, then anxiety, anger, and feelings of insecurity became more pronounced among all the family members. The author reports that young people will generally believe what they want to believe, for to confront the implications of a deteriorating disability is simply too threatening. Power does not provide support for his view concerning adolescents believing what they want to believe. It may well be that adolescents are engaged in emotion-focused coping methods, which includes positive denial (Lazarus, 1984).

Family expectations with regard to the performance of duties in the home were shown to have implications as well. It was found that the higher the expectations of the family members, the more the ill parent was willing to participate in social activities and household chores. These activities facilitated the affected parents’ greater participation in the life of the children.

Family acceptance refers to the family’s positive adjustment to the disability, and is shown by methods of coping and the pattern of activities in the home. The achievement of family acceptance is usually precipitated by the family’s understanding of the disease and its disabling
aspects, as well as by positive expectations for each other. An additional factor is family members sharing with each other their own feelings of loss caused by the disease itself. Where adolescents were adapting poorly to the disease impact it usually followed that family members were seeing the illness situation from different and conflicting views.

The study indicated that the nature of the parent’s chronic illness as well as the family dynamics could influence the adjustment pattern of adolescents. Power claims that while young people can survive the impact of HD and MS on the family, the inheritability aspect of HD can present extreme difficulties for the coping process. Regardless of the interviewing techniques used, Power’s study does point to the importance of understanding chronic illness from the context of the family, and he contributes to an understanding of the adolescent experience of being at risk for HD.

Korer and Fitzsimmons (1987) conducted a semi-structured interview of fifty adolescents at risk of developing Huntington’s Disease in order to explore their attitudes toward the condition, and to establish how helpful genetic counselling would be for them. They were concerned that young people may be left in ignorance about HD: the findings of previous studies showed that parents of young people at risk for HD felt burdened by the responsibility of giving genetic information to their children, and found the responsibility too great to cope with alone (Korer & Fitzsimmons, 1985; Tyler & Harper, 1983; Barette & Marsden, 1979).

Two groups of young people were selected. Group one was composed of those with a parent who had died or who was currently suffering from HD. This group had a 50% chance of developing the disorder. Group two was composed of young people who had a 25% chance of developing the disorder.8 It was considered likely that those in group one would have additional psychosocial problems, and that group two individuals would be an interesting comparison group

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8 This group consisted of adolescents who had a grandparent who had died or who was currently suffering from the disease; however, if a parent developed the disorder their risk would automatically rise to 50%.
given that they presumably had a less disrupted family life and therefore more closely resembled “ordinary” young people.

The researchers state that it was much more difficult to introduce the subject of Huntington’s Disease with the parents in group two than in group one. To the group one parents the facts were obvious, and individual families welcomed the contact of the research interview. The group two parents, however, were much more wary and often required several visits before permission was given to contact their children.

The results of this study were illuminating. More than half the young people in group one had left school without graduation or training compared with less than one-quarter in group two. Nearly 50% of group one and 86% of group two had not told any of their friends about their own risk for HD. This suggests that a fairly high proportion were keeping a significant “secret” that they felt unable to share with any friend, no matter how close. The authors questioned the adolescents on both their factual and experiential knowledge of HD. Most people at risk for HD are what Nancy Wexler (1979) calls “disease wise,” that is, from a very early age they know what the disease is called and some basic facts about its inheritability (it’s “in the blood,” it’s “passed down in families”). The study found that most people in each group had some understanding that the disease is passed down from one generation to another; however, many of the adolescents had picked up erroneous information. It was found that if the parents’ information was inaccurate the erroneous information would be passed on to the children.

Like the earlier studies, Korer and Fitzsimmons did not ask adolescents how they coped with being at risk for HD, or what they imagined when they considered their future. However, there were some spontaneous responses from the adolescents about being at risk, with the interviewer citing three respondents experiencing severe psychological difficulties.

In the discussion section of the research, the interviewer suggests that the adolescents in the study seemed to be better able to cope with “the dilemmas engendered by HD than did their
parents." They also reported that interviews with the parents revealed that they would relinquish the responsibility of telling their children the facts about HD if they knew their children were receiving skilful genetic counselling. The adolescents in the study who had received genetic counselling reported that they had found it helpful. It was found that the young people were able to discuss HD and its implications in a "rational and thoughtful manner," and that there was "no evidence to support the supposition that young people are too emotionally volatile to cope with information about HD" (1987:532). Nevertheless, the authors conclude that they did not feel it was "appropriate to enquire directly how the young people interviewed coped with being at-risk and with the prospect of what might lie ahead" (1987:532). Research into the experience of children at risk was not attempted, apparently to avoid confronting adolescents with uncertainty or unpleasant and upsetting futures.

While disturbing adolescents for the sake of research is a valid concern, the discomfort of researchers may also be a significant factor that continues to influence the direction of research into Huntington’s Disease. As Korer and Fitzsimmons (1985:596) point out, “No one can remove the unpleasant facts of the disease or eliminate uncertainty and insecurity.” They emphasize that “work with these families” was bound to be “difficult and distressing.”

**Development of Predictive Testing for Huntington’s Disease**

With the advent in 1986 (one year after the Korer and Fitzsimmons study) of presymptomatic DNA testing using genetic linkage analysis, and with the identification of the HD gene mutation in 1993, people at risk for HD have been able to receive predictive test results. They learn that they have either inherited or not inherited the specific genetic mutation associated with HD. For many patients receiving this information the question changes from

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9 The linkage test is still probabilistic; the direct test is more definitive.
whether they will develop HD to when will it manifest (Burgess & Hayden, 1996). Test results
do not reveal when gene carriers will develop onset, nor is it possible to predict the probable
severity of the disease at onset.

The increased accuracy and certainty of the direct test imposes a new burden of
responsibility on clinicians and researchers. Frankl (1978) contends that individuals need a
vision of the future to thrive. For those at risk for HD, predictive testing has profound
implications, as the results of direct testing could significantly alter one’s notion of the future. As
Mattsson and Amlqvist (1991:26) state:

It is a serious exhortation to those who accelerate the technical development not
to ignore the ethical consequences of such a test. When reliability of the test and
technical perfection is 100% all hopes of not being a victim are dashed to the
ground. Without such hope the human consequences of suffering, fear and
irresoluteness may be high.

Such concerns are reflected in researchers’ shift from the question of whether or not to
offer predictive testing to how best to continue to offer it (Cox et al., 1995). Pre-test counselling
protocols have been established to “set a standard of good practice for preparing and supporting
persons who request predictive testing” (Craufurd & Tyler, 1992: 916). Numerous studies
conducted throughout the world have examined the psychological effects of DNA testing (Bloch
et al., 1992; Cordori & Brandt, 1994a, 1994b; Evers-Kiebooms et al., 1990; Huggins et al., 1992;
Quaid, 1993; Quaid & Wesson, 1995; Tibben et al., 1993a; Wiggins et al., 1992). Generally, the
results demonstrate that individuals who are tested find relief from prior psychological distress
and, in fact, benefit psychologically from testing.

As the literature reveals, less attention has been devoted to investigating the familial and
social implications of predictive testing. Although it is increasingly recognized that Huntington’s
Disease is a family problem, few multi-disciplinary research approaches exist that combine the
clinical, counselling, and social science expertise necessary to undertake this type of
investigation. Further, as demonstrated in Korer and Fitzsimmons’ (1987) study of adolescents at risk for HD, some social and medical service providers may be reluctant to involve family members in research that could stir up thoughts and feelings that lead to increased psychological distress. The psychological support potentially needed by families may increase demands on an already overburdened counselling infrastructure.

**Adolescents and Predictive Testing**

What if children under the age of eighteen request to know of their risk status? Internationally accepted guidelines have been issued to “protect at-risk persons” and “to assist clinicians, geneticist, and ethical committees as well as lay organizations to resolve difficulties arising from the application of the test” (International Huntington Association and the World Federation of Neurology Research Group of HD, 1994:555). One of these guidelines precludes testing of children under the age of eighteen on the grounds that such profound decisions should not be made until the child concerned has reached legal age and can make that decision independently. The exclusion of minors from predictive testing for Huntington’s Disease is rationalized on the grounds of the lack of clear benefits of testing at this age, the possibility of third-party pressure in the request, and the minor’s inability to make an informed choice (Harper & Clarke, 1990). It is necessary to look carefully at the ground rules surrounding predictive testing as adolescents are beginning to request the test for Huntington’s Disease (Binedell et al., 1996).

Van der Steenstraten et al. (1994) studied members of the Dutch Huntington’s Association who were at 50% risk in order to determine the reasons why some of them chose not to participate in predictive testing programs. The group of thirty-four non-participants completed the same baseline psychological questionnaires as did those who chose to participate in the testing program. Non-participants in this study did not believe that the predictive test for HD
would be a realistic option for improving their quality of life. It was also discovered that in comparison with participants, non-participants had a significantly more pessimistic outlook on themselves and their futures. The non-participants stated that a favourable result would reduce problems for their children but not for themselves. An important finding in this study was the age at which the non-participants learned about being at risk for HD: The non-participants learned about their risk during adolescence (mean age 15.6 years), whereas the participants did so in adulthood (mean age 22.7).

The researchers speculate that being informed of the disease and of personal risk at different developmental stages of life could be a possible explanation for the different attitudes exhibited toward the predictive test. They speculate that the process of individuation and separation (Blos, 1982) may have been influenced and burdened by the "fearsome idea of an HD-doomed adulthood and the expected loyalty toward an affected parent (1982:623)." Further, feelings of guilt, anxiety, and anger may also contribute to an increase in pessimistic expectations with regard to the future. They speculate that participants learned about HD when most of their basic choices in life regarding education, partnership, and career had been decided, and then their life-concept was interrupted by the knowledge of their 50% risk status for HD. This knowledge may have disturbed future expectations, but participants might have wanted to restore balance and increase self-control by undergoing predictive testing. It is further speculated that non-participants may have tried to control their future by coping mechanisms involving denial and minimization. The non-participants' reluctance to take the test is viewed by the researchers as a reflection of harm-avoidance behaviour. They believe that this coping mechanism may remain unnoticed by professional caregivers. The findings suggest that future research on the psychological effects of predictive testing should investigate the development of personality, with the inclusion of coping styles and psychological defence mechanisms in
individuals confronted in childhood and adolescence with a personal risk for late-onset genetic disease.

The Unanswered Questions

It appears that there is an implicit belief guiding the inquiry into the effects of predictive testing: knowledge is intrinsically good and it will enable people to make more informed decisions. It is what Kenen (1996:1545) calls “the diagnostic invitation and the gift of knowing.” Knowledge is equated with empowerment, yet when diagnosis merely reaffirms risk and offers no cure in the near future is it beneficial? An at-risk individual may welcome this gift of knowing, feel ambivalent towards it, or reject it. Does the rejection of predictive testing imply that the non-participant has suffered from an interrupted developmental stage and has adapted using coping mechanisms such as denial and minimization? I contend that the decision to take or reject predictive testing will depend on a wide variety of circumstances and individual values that are yet to be studied.

Prior to linkage analysis and the identification of the HD gene mutation, research into Huntington’s Disease had just begun to explore the familial and social implications of the disease in the lives of the affected person and their family members. As noted previously, a few studies acknowledged the dilemmas faced by children within the HD family, and some addressed the adolescent experience in an HD family (Power, 1984; Korer & Fitzsimmons, 1987). With the advent of new diagnostic technology, the focus shifted from assessing whether predictive testing was beneficial and justified as a service to predicting and reducing associated risks. Adolescent testing for HD is prohibited – without addressing the possibility that testing might be beneficial for particular individuals.

Current research is now exploring the psychological benefits of testing, and the effects of testing on the affected individual and on the family members (Tibben et al., 1993a, 1993b, 58
In recent years, adolescents have come forward and requested testing in order to determine their risk status (Binedell et al., 1996). This has lead to a debate in the medical community concerning the ethical, legal, and psychological implications of testing healthy children and adolescents for disorders such as Huntington’s Disease (Wertz et al., 1994).

“As technological knowledge expands, so does the power of the geneticist to influence the lives of clients who seek counselling about their own genetic status or the status of their children” (Pelias, 1991:347). It is my contention that the rapid advancement of new technologies in the field of medical genetics has caused the medical community to shift its focus prematurely without adequate information on the impact of the disease in HD families. Researchers and medical care providers grapple with issues of predictive testing and its effects on individuals and families without having gathered information about how the presence of the disease affects the attitudes, beliefs, and behaviours of the family. In other words, the effect of predictive testing on at-risk persons and families cannot be distinguished from the effect of being at-risk or a member of an at-risk family.

It seems probable that children of HD-affected parents grow up under stressful psychological and social conditions. It is imperative to begin the groundwork necessary to determine how the presence of Huntington’s Disease in a family affects the at-risk adolescent. What are the experiences of adolescents whose parents are at risk for HD? What are the experiences of adolescents whose parents are tested for HD? What is the potential impact of the adolescents’ at-risk status on their view of the future? An understanding of these issues will guide families and clinicians who want to support adolescents. It could also provide the basis for an evaluation of whether predictive testing might sometimes be beneficial for adolescents, and for a comparison of the effects of predictive testing as distinct from being at risk for HD.
CHAPTER III

METHODS AND PROCEDURES

Phenomenology requires a kind of withdrawal from the world, and a willingness to lay aside existing theories and beliefs. This is risky, and takes an act of courage. It can be viewed as a journey during which one leaves familiar places and then returns and sees these places in a fresh light (McLeod, 2001:37).

Purpose and Outline of Chapter

This chapter begins with an overview of phenomenological method that is intended to assist the reader in understanding why this particular qualitative method was judged to be the most effective research tool for illuminating the experience of adolescents at risk for Huntington’s Disease. The chapter goes on to explore the possible biases of the researcher, and to delineate Gottlieb’s (1994) decision-making model, which was used as a means for assessing the potential for confusion that might arise from the various roles that I play in the HD community outside of this research.

A review of the theoretical underpinnings that fuelled this research and influenced the choice of the phenomenological method will be presented. A section on research design delineates the procedure that I followed, and covers recruitment and participation rate, ethical considerations, the use of the “lifeline” as a research technique, fieldwork and other sources of data, and the process of analysis. The last section of the chapter examines the limitations of the study by discussing the validity and reliability of the research process.

Overview of Phenomenological Method

Phenomenological psychology has evolved from the philosophical theories of existentialism and phenomenology, especially as presented in the work of Husserl (1970). Phenomenology aims for “a deeper and fuller understanding of human existence, ourselves, and
others” (Valle et al., 1989:16). Phenomenological methods were developed to describe and understand phenomena (e.g., events, occurrences, experiences) of the “life-world”; the world of experience as actually lived rather than as conceptualized, categorized or theorized (Van Manen, 1984:27). To this end, it is a methodology that helps counter the prevailing behaviourist and cognitive emphasis in psychological inquiry. As several writers have contended, traditional experimental modes of inquiry, because they are grounded in scientific positivism, fail to explain many of the realities of human experience (Giorgi, 1985; Polkinghorne, 1989; Valle et al., 1989). The natural scientific model that explores outwardly observable human behaviour may be inadequate for “the inward, unobservable side of others; that is, their thoughts, emotions, and sensations, commonly referred to as their private world of experience” (Valle et al., 1989:3).

To the existential-phenomenological psychologist “only that which has its base in naïve experience is real and, therefore, only that which is revealed or disclosed as pure phenomena is worthy of attention” (Valle et al., 1989:13) whereas the natural scientist equates objectivity with distance from the phenomenon. The phenomenological approach concentrates not on describing physical objects, but on describing experience. Phenomenological psychology differs from mainstream psychology by holding that human behaviour is “an expression of meaningful experience rather than a mechanically learned response to stimuli” (Polkinghorne, 1989:41). Moreover, phenomenological researchers accept that the “truth” about any investigated phenomenon emerges not as one absolute and objective view, but as a composite picture of how the individual(s) who have experienced that phenomenon perceive or understand it (Bogdan & Taylor, 1975). Thus, phenomenology views people as “unique,” unlike the natural science approach that “aims at knowledge that is generalizable to everybody” (Van Manen, 1990:6). Meaning is, therefore, considered distinctly personal, as each person experiences life in a unique way.
According to Giorgi (1985), the purpose of phenomenological research is to undertake “direct analysis of the psychological meaning of naïve descriptions of personal experiences provided by individuals from all walks of life in situations that are easily recognizable as belonging to everyday life” (1985:1). In order to understand and describe a phenomenon in all its diverse appearances, the existential-phenomenologist seeks to reveal the structure of the phenomenon – that is, its meaning. To reveal this structure, the existential-phenomenological psychologist must ask *what* not *why*.

That is, he or she seeks to understand phenomena in their perceived immediacy and is not concerned with explaining, predicting, or controlling them – the question, *why?* is not asked as this question implies an underlying causal view of the world. (Valle et al., 1989:13)

Phenomenological reflection is retrospective, a “reflection on experience that is already passed or lived through” (Van Manen, 1990:10). Participants in phenomenological research should be able to recollect their experience as they lived it. To do a phenomenological study, Van Manen (1984:7) stated,

The experience must be recalled in such a way that the essential aspects, the meaning structure of this experience as lived through, are brought back, as it were, and in such a way that we recognize this description as a possible human experience, which means as a possible interpretation of that experience.

However, the problem is that we often know too much about the phenomenon we are investigating and our assumptions “predispose us to interpret the nature of the phenomenon before we have even come to grips with the significance of the phenomenological question” (Van Manen, 1990:46).

Phenomenological researchers concerned with obtaining a truthful account of lived experience must take care “to avoid leading the person, imposing biases, or planting elements of one’s own” (Cochran & Claspell, 1987:42). They do this by making their assumptions explicit prior to engaging in research. That is, they bracket – or put out of play – everything they “know”
(personal and cultural assumptions) about the specific human experience they are investigating in order to prevent their assumptions from influencing their research (Van Manen, 1990:47).

**Situating Self in Research**

**Multiple Roles**

I have been engaged in the Huntington’s Disease community as both a volunteer and a professional in two different settings. I had a two-year internship as a therapist in the Department of Medical Genetics in Health Sciences Centre at the University of British Columbia, and met many families with HD through my role at the clinic. I also volunteered at the yearly retreats sponsored by the Huntington Society of B.C., where my role became identified with therapy. I created groups that were psycho-educational, and also spent one-on-one time assisting retreat participants who were struggling with issues related to HD. I realized that my capacity as a therapist was a factor that allowed some adolescents and parents to feel more comfortable with their involvement in the research that forms the basis of this paper. I also came to realize that my therapeutic skills might be necessary and beneficial. In my work with adolescents I have found that the interview process – the telling of the story – can result in the remembrance of traumatic events that can lead to trauma symptoms. Traumatic affect containment techniques are then required to ensure the well-being of the individual.\(^{10}\) I have had occasion over the course of my years as a therapist to use such techniques. Consequently, I felt more comfortable engaging in the research knowing that I had three decades of therapeutic skills to assist me.

\(^{10}\) Traumatic experiences have often been encoded differently and often at a nonverbal level. Traumatic affect containment techniques allow the therapist to successfully help a traumatized individual with trauma-related affect, defences, and dissociative states (Bromberg, 1998).
Just as the therapist’s counter transference operates as a rich source of information about the client and the state of the therapeutic relationship, the feelings I experienced as a researcher have given me clues as to what is most important in the research situation. I believe that qualitative research and therapy have a number of aspects in common. What was most obvious for me was that each of these modalities seeks to empower the people who participate in the process (Vesper & Brock, 1991; Wolcott, 1994). My colleagues helped me to appreciate how qualitative research can be viewed as a vehicle of “social transformation” (Fine, 1992, p. 209).

A primary focus in my work as a therapist is to help empower my clients so that they will be able to bring their own skills and competence to their personal lives; thus, the association of qualitative research and therapy can be viewed as closely aligned, as each has the goal of enabling others to act from their own power (Bourdeau, 2000).

Qualitative research and therapy have many structural similarities as well. Both are involved in a process that requires interviewing sessions that can be quite intense; both can create a climate wherein a person divulges intimate and personal information to another person whose role it is to listen and ask probing questions. Depending upon the length of the interviews and the duration of the research, it is possible that some qualitative research can create more depth of therapeutic process than therapy itself.

A number of factors became important for my research with the adolescents. The first and most important issue for me was that of power. The structural similarity of therapy and qualitative research involving interviewing creates an inherent power variance. In fact, some feminist researchers (Allen & Baber, 1992; Harding, 1987) recommend that researchers study

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11 I attended weekly research meetings at the Centre of Applied Ethics at the University of British Columbia. It was an interdisciplinary team that supported qualitative research in genetics and ethics and created a venue for discussions across disciplines on research topics of the group.
only those at the same or higher level of social power. La Rossa et al. suggest that there is a "relative powerlessness of the subject vis-à-vis the researcher" (1981:306) inherent in the structural nature of qualitative research. This inherent power variance and the issue of subject compliance became exacerbated by the fact that my research participants were adolescents.

The Tri-Council Policy Statement: Ethical Conduct for Research Involving Humans has developed guidelines that describe the policies of the Medical Research Council (MRC), the Natural Sciences and Engineering Research Council (NSERC), and the Social Sciences and Humanities Research Council (SSHRC) (1999). These guidelines assist in promoting and implementing ethical standards in research involving human subjects. The guidelines assisted me in the over-all conduct of the research but Gottlieb’s (1993) decision-making model addressed directly the dual relationship dilemma I experienced in this research.

In order to keep in focus this potential differential in roles and the possible consequence of participant compliance, I employed the decision-making model developed by Michael Gottlieb (1993). This model addresses the dual relationship dilemmas encountered by therapists. It provided a means to bring into awareness the multiple roles that I play in the HD community and with my co-participants, and it helped me to recognize when my dual role caused me to cross the boundary of researcher and in some way affect participant compliance.

The Gottlieb Decision-making Model

Gottlieb’s (1993) decision-making model classifies three dimensions of dual relationship dilemmas, each containing three levels of intensity. The first dimension is power, which varies from low to mid-range to high. The levels of power are based on the amount of vulnerability required of the participant/client, and the amount of influence maintained by the researcher/therapist. A relationship that is low power would be one where the researcher/therapist and the participant would be considered peers. Given that the participants in
this study are adolescents, my relationship with the participants would be classified as a “high power relationship.”

The second dimension is duration of the relationship, which is measured from brief duration to intermediate to long. Contact is on a continuum from few contacts over a brief amount of time, to continuous contact over a long period of time. Given that I am still in contact with some of the participants in this study through one-on-one meetings, phone calls, and e-mail I again would be placed into the more intense realm of “long duration.” The final dimension is clarity of the termination, which ranges from specific to indefinite. With specific termination, there is a date when the professional relationship will end. In order for me to have conducted this study, I could not be a stranger in the community. I was engaged in the community and could be viewed as a participant observer acting within the community. In my case, the indefinite nature of termination due to the role I play outside of research and within the HD community created a lack of clarity about the end of the relationship, with no time limit on the professional nature of the relationship.

When making decisions based on this model, the therapist/researcher tries to place the relationship in terms of the three dimensions from the point of view of the participant/client. Applying this model to my dual role in this research helped me appreciate my position as having the potential for harm. There was a high power differential. I am still in contact with many of the participants and their families, and, therefore, the duration and indefinite termination of a relationship with the participants required me to maintain a professional relationship and to avoid a non-professional relationship. For example, two of the adolescents in this study decided to engage in predictive testing and have since solicited my professional support. I have engaged in individual and family counselling with these adolescents and continue to assist them in coping with the implications of their test results. In this situation, the decision-making model has provided me with a means to navigate through the multiple roles I experienced as researcher,
therapist, and volunteer, as well as the power differential that was inherent in my dual role as therapist and researcher.

The interplay between these roles shaped and influenced the conversations I had with the adolescents in other ways. As a therapist, I am often concerned with how a client perceives support in their personal and social worlds, and with enabling them to find the support they need during a particular life crisis. Consequently, many of the conversations that were a part of this research were about "how" the adolescents experienced being at risk rather than "what" they experienced being at risk. It is through exploring "how" that I learn about the context of their everyday experience and the kind of relationships and activities that they perceive as supportive. The stories that the adolescents told confirmed this assumption; they often involved discussions around how the adolescents coped in everyday life and how they perceived support or lack of support. An adolescent's experience with her grandfather in the hospital is an example of this nuance in perspective: The "what" of her experience would be that her grandfather was sick and she attended the hospital with her mother. It was during this time that she first learned that her grandfather had a disease called Huntington's. The "how" of the experience was that she played dolls with her grandfather and he allowed her to push him in a wheelchair through the hospital gardens. What she experienced was her grandfather in the hospital, and a name – HD – given to his illness. How she experienced her grandfather was through the eyes of a child who had the attention of a grandfather who was a captive playmate and who became part of a joyful memory of childhood. Gottlieb's model helped me to ask the questions that were pertinent to the research questions and to refrain from therapeutic questions that may have been inappropriate for the role of researcher and the aims of the research.

Realization of another way in which I was influenced came when I recognized that the process of intuition, learning, and understanding that delineates the adolescent's path of learning about Huntington's Disease mirrored my own experience as a student/researcher. I began this
research based on some intuitions and experiences I had with adolescents in the HD community. As I progressed with the research, I began to see that there were similarities between how I learned about the adolescent experience of HD and how the adolescents themselves experienced learning about HD. Much like the adolescents in this research, I moved from intuition, factual knowledge, and experiential knowledge to the insight I present in my research.

**Methodological Approach**

This dissertation is about the experience of adolescents at risk for Huntington’s Disease. I pursued a qualitative study using a phenomenological approach. As discussed in Chapter II, an initial survey of the literature produced little evidence of empirical research work regarding the experience of the at-risk adolescent whose parent is affected by HD. Chronic illness and its significant impact on the family is well documented; however, even a careful reading of such research will not reveal an adequate portrayal of the experience of children in a family with an HD-affected parent.

**Theoretical Underpinnings**

The research continually presented new views and perspectives as the interviews progressed. My challenge was to stay open to the many different visions, to resist my desire for closure – that impatient desire for the quick “right answer.” Qualitative research provides for a range of feelings – including the desire for closure – and recognizes that a wide range of emotions are intrinsic to good practice rather than indicative of personal shortcomings (McLeod, 2001). It also allowed for an approach that gave the adolescents an opportunity to express in their own words and in their own style what the experience of being at risk for HD meant to them within the context of their own family history, stage of development, and awareness of the disease, while still allowing me to attend to themes that might arise from their different visions.
Family therapy has taught me that the best way to understand phenomena is to view it in context. I became involved in the Huntington’s community, created an adolescent support group, and attended weeklong retreats for people with HD on a yearly basis. I designed support groups for these retreats that were intended to assist individuals with the ongoing challenges presented by being a person with HD symptoms. I became immersed in the phenomenon of HD, and involved in the culture and organizations. I learned many valuable lessons from immersing myself in the HD culture. I learned about the need for flexibility in my exchanges with retreat participants and support group attendees.\textsuperscript{12} I also learned that I was a guest in their experience of HD, and that as such, I needed to respect their experiences and recognize that my inclusion in their world was a privilege extended to me.

I then began to deconstruct my own epistemological and ontological assumptions in order to choose the appropriate research methodology. It was in phenomenology’s interpretive methodology that I found support for my experience. In phenomenology, the epistemological assumption is that the researcher interacts with the participants, and the researcher’s role is to understand the participants’ interpretation of events rather than the events themselves. Pure phenomenological research seeks essentially to describe rather than to explain, and to start from a perspective free from hypotheses or preconceptions (Husserl, 1970). More recent humanist and feminist researchers refute the possibility of starting without preconceptions or bias, and emphasize the importance of making clear how interpretations and meanings have been placed on findings, as well as making the researcher visible in the “frame” of the research as an interested and subjective actor rather than a detached and impartial observer (Plummer, 1983; Stanley & Wise, 1993).

\textsuperscript{12} Huntington’s Disease has multidimensional symptom severity. Although there are symptoms that occur across individuals who have HD, it manifests differently in each individual.
The aim of phenomenology is to produce an exhaustive description of the phenomena of everyday experience in order to understand the essential structures of the “thing itself” – the phenomenon (Moran, 2000). From a phenomenological point of view, we are not primarily interested in the subjective experiences of the participants for the sake of being able to report on how something is seen from their particular view; rather, the aim is to collect examples of possible experiences in order to reflect on the meanings that are inherent in them (Van Manen, 1998).

As I examined my ontological assumptions about reality, it became clear that I could not conduct research that did not acknowledge that each of us experiences reality from our own point of view, and, therefore, we cannot escape our subjective experience of reality. Based on my belief about reality, I was opposed to methods that attempted to aggregate across individuals, as I experience each individual as unique. In fact, I as a researcher am also a unique individual; my research is influenced by my own perceptions and ways of conducting the research. An example of this bias is reflected in my experience of mothering six children through adolescence. It was a remarkable learning experience for me, and I emerged from that decade of my life with an incredible appreciation for the resilience and creativity of youth. It would be impossible to separate my positive affect regarding the capacity of adolescents to cope from my role as researcher with the adolescents involved in this study.

**Research Design**

As demonstrated in the literature review, the methodology utilized in research on HD has been primarily deductive in orientation; it has focused on people at risk as “patients,” thus reflecting a medical rather than a social focus. Conrad recognizes the importance of a social focus in research directed at chronic illnesses like Huntington’s Disease.
Whether and to what degree we are able to make a contribution to clinical medicine depends in part on the rigor and insight of our research, in part on our ability to persuade our medical colleagues of the significance of our analysis, and in part on patients and physicians recognizing that the major issues in managing long-term chronic illness are probably more likely to be social than medical (1990:1257).

Also relevant is the fact that where research addresses adolescents and HD it may not realistically reflect the adolescent experience, as it is parents or other family members who supply much of the information concerning the adolescent experience. I believe, however, that in order to elucidate the adolescent experience of living with the risk of and in spite of HD, it is imperative that the adolescents themselves be the focus of the investigation. In this regard, a qualitative research design is particularly well suited to studying and understanding the subjective aspects of being at risk for HD.

**Recruitment and Participation Rate**

The participants eligible for this study were adolescents between the ages of thirteen and nineteen who were at 50% risk for Huntington's Disease. Risk status was conferred with a parent receiving a positive predictive test result for HD, and/or a parent having symptomatology associated with HD.

When approached about this study the parents and the adolescents consistently demonstrated a willingness to participate. Only one adolescent did not participate in the study after I had received parental consent. His father informed me that his son was having difficulty in school and at home. The father was a single parent and was uncertain that his son would remain with him. In fact, when I tried to make arrangements to talk with the adolescent, he could never

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13 He did eventually run away from home and moved to another area of the province.
be found as he lived in several of his friend’s homes in a sporadic manner. It was probable that he would not have been able to complete the interviewing process.

A number of factors may have contributed to the high level of participation: I was able to have personal contact with eight of the nine adolescents and/or parents prior to recruitment. This gave the adolescents and their parents a chance to get to know me and to get a sense of how I was going to conduct the interviews. The remaining adolescent was recruited through a social worker with the HD Society. The recruitment process provided some time prior to the interviews for the adolescents as well as their parents to consider the research more fully, and to identify areas about which they were not clear, or areas where they had some questions. Additionally, all of the families of the adolescents I interviewed had been involved with the HD Society and had learned of my work through the society or the Huntington’s Disease Clinic at the Department of Medical Genetics in the Health Sciences Centre at the University of British Columbia (UBC). Eight of the adolescents’ parents had met me either through the Huntington’s Disease Clinic at UBC or at provincial or national meetings. I was known within the community, and I worked at a clinic where people, in general, were appreciative of the support and counselling provided. The endorsement of the society and the clinic helped to establish a level of trust and support for me in my role as researcher as well as my role as a researcher/therapist. Most importantly, the interviews provided an opportunity for the adolescents to talk about living in a family with Huntington’s Disease. For all of the adolescents involved in this research, it was through the interview process that their stories were told for the first time. All of the participants reported that the opportunity to tell their story was significant for them.

The participants were recruited in several ways. Recruitment was undertaken through the Huntington’s Disease Clinic at the Department of Medical Genetics in the Health Sciences Centre at the University of British Columbia where I was a therapist. The recruitment was not limited to British Columbia. Based on my attendance at the national conferences of the
Huntington Society for Canada, my participants were recruited from three provinces across Canada. I approached parents of at-risk adolescents and explained the study in detail to them. Participants were also recruited through the British Columbia chapter of the Huntington’s Disease Society where I was running an adolescent support group. The social worker for the society was helpful in contacting families for me. In each case I gave the parents a detailed explanation of the study. They were asked if they would agree to have their children participate. If parental permission was obtained, I met with the adolescent and explained the study in terms appropriate to their level of development, which was determined based on their age and my experience in working with adolescents. The younger the adolescent, the more I would explain the process and the more I would verify their understanding by asking them to summarize what they understood about the study. This system of detailed discussion and adolescent summary helped to ensure that the adolescents were clear about their consent to participate regardless of their age. Prior to engaging in the interview process, the adolescents and their parents were given an opportunity to discuss any questions or concerns with me, either by phone or in a meeting.

Table 1 illustrates the demographics of the adolescents.
TABLE 1
CO-PARTICIPANTS’ DEMOGRAPHICS

<table>
<thead>
<tr>
<th></th>
<th>Stephanie</th>
<th>Carla</th>
<th>Marilyn</th>
<th>John</th>
<th>Daphne</th>
<th>Emma</th>
<th>Tessa</th>
<th>Margaret</th>
<th>Kathy</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age</td>
<td>14</td>
<td>15</td>
<td>16</td>
<td>18</td>
<td>19</td>
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<td>13</td>
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</tr>
<tr>
<td>Rural/Urban</td>
<td>R</td>
<td>U</td>
<td>U</td>
<td>U</td>
<td>U</td>
<td>R</td>
<td>U</td>
<td>U</td>
<td>U</td>
</tr>
<tr>
<td>Affected Parent</td>
<td>Dad</td>
<td>Mom</td>
<td>Dad</td>
<td>Dad</td>
<td>Dad</td>
<td>Mom</td>
<td>Mom</td>
<td>Mom</td>
<td></td>
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<td>4th of 4</td>
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<td>2nd of 4</td>
</tr>
<tr>
<td></td>
<td>Brother too young for study</td>
<td>Siblings</td>
<td>Brother too young for study</td>
<td>Siblings</td>
<td>Eldest sibling too old for study</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Ethical Considerations

There were essentially two levels of concern with regard to the ethical motivations of this research. The first motivation derived from the fact that research on Huntington’s Disease lacks first-hand accounts of the experiences of children and adolescents that are presented in their own voices. The second motivation comes from the need to ensure that the conduct of this particular research provided an opportunity for adolescents to present themselves in their own way so as to help illuminate their experience.

There is wide variation in the literature on children and their experiences of family and home. Burman (1994) states that there exists conceptual frameworks that are imprecise and contain many hidden assumptions. Above all, the reflections of children themselves are conspicuously absent from the literature, and children are seldom given an opportunity to relate their own experiences - they come to be defined in large part by adults (Moore et al., 1996).

Leach (1994) takes this one step further by suggesting that children are a minority group subject to discrimination. The rights of children came to international attention in the 1990s. Franklin (1995) provides a comprehensive view of the comparative policy and practice regarding
children's rights and the need for a greater respect for children in matters that apply directly to them. In fact, the United Nation's Convention on the Rights of the Child states that one of the most important rights of a child is the right to be heard (UNICEF, 1995). Moore et al. (1996:2) contends that "children's rights to have their reflections taken seriously is part of their entitlement to a full and decent life."

The absence of the child's voice is also apparent in the literature on Huntington's Disease. One of the motivations for the methodology used in this research was to give adolescents an opportunity to present themselves in their own way, and thus to provide information that truly reflects the adolescent experience of living with HD.

This research project received ethical approval from the University of British Columbia's Office of Research Services and Administration Behavioural Research Ethics Board. Full provision for informed consent, confidentiality, and security of data were included in the application for ethical review. Copies of the letter of introduction and consent form are included in Appendix II.

There were certain aspects of the research design that were salient to "doing research on sensitive topics" (Lee, 1993) and that required careful consideration. The ethical considerations in this research were validity of the adolescent consent, confidentiality and clear understanding of the regulations regarding report of abuse, and possible harm that may be incurred from recounting traumatic experiences. Because my participants were adolescents, it became necessary to take special measures to ensure that the adolescents were engaged in meaningful, informed consent. The procedure by which consent was obtained, as well the consent form itself, took into consideration the potential for a lack of comprehension on the part of the adolescents of what volunteering implied. In the consent form, and in my verbal disclosure concerning the research, I emphasized that participation was understood as a contribution, that there was no obligation, and that participants could withdraw at any time. I repeated this orally at every
contact. The parents and the adolescents in this study seemed to understand the research process, and the adolescents were actually eager to engage in the process.

I spoke to both the adolescents and the parents about the potential risks posed by participation in the study, including the possibility that the interview process might give rise to thoughts or feelings that had not been openly discussed by family members. I also felt that it was possible that suspicion of child abuse occurring in the family or in the social world of the family and adolescent might be revealed in the interviews. I told the participants and their parents about this possibility both in the consent forms and verbally. Before the interviews began I informed the participants of the legal responsibilities of the researcher in the event that information regarding self-harm, abuse, or harm to others was given. I explained what the provincial reporting mechanisms were and my obligation to report. I emphasized that there was no direct benefit to be derived from participating in the study.\footnote{14}

Although the adolescents and their parents knew that I was a therapist, I did not interview any adolescent that I had previously engaged in therapy. Two of the adolescents had attended the adolescent support group. The group was designed to be educational and provide an opportunity for adolescents to speak to one another and potentially normalize their experience of growing up in a family with Huntington's Disease. It was not designed to be a therapeutic group although there was the opportunity for a therapeutic benefit by adolescents engaging with other adolescents who came from families that may have had some of the same challenges that had not been discussed inside or outside of the family. Additionally, I assured both the parents and the adolescents that genetic and family counselling was available should they feel the need for additional information or support.

\footnote{14 In one instance an adolescent did benefit directly from the study. Carla revealed engaging in suicidal ideation in the interview. I managed this disclosure by exploring the ideation and securing counselling for Carla (see page 206).}
Confidentiality was an important issue for the adolescents. As stated, each of the participants was telling their story about living at risk for HD in an HD family for the first time. They wanted to be free to talk about their experiences without worrying about how family members would react to the mention of certain subjects or incidents. I dealt with this concern by describing how the interview tapes would be maintained. I explained that individual ID numbers, not names, would identify the tapes, and that all computer files would be kept nameless. I assured the adolescents that tapes and transcriptions of the interviews would be kept in a locked cabinet, to be accessed only by myself. The Huntington Disease community is a small community and the participants and their families could be easily identified. In every case, parental permission was given to allow me full license to use the accounts that the adolescents provided in the research. I assured the participants and their parents that their names and any personal information that might identify them would be altered in the final research documents. The identification of the families was minimized by changes to inconsequential demographics and by the geographical spread over three provinces. Although permission was given by the parents to use the accounts and measures were taken to insure anonymity, it remains a possibility that the parents may identify their child’s story and become upset upon seeing what the adolescent said. My continuing involvement with the Huntington’s community means that I am available for the parents, adolescents or other community members who may have concerns about what they read.

I also let the adolescents decide where they wanted to be interviewed. Six of the participants chose to be interviewed away from their homes. Three of the adolescents were interviewed at home: one adolescent had me attend when no adults were present, and two of the adolescents had me conduct the interview in a part of the house removed from the activities of home life. I conducted an interview on an apartment porch, and another in the basement of the family home. Additionally, I reminded each study participant that they could request to have the
tape recorder turned off at any time to keep certain information “off the record.” I also reiterated that they were free to disengage from the research project at any time.

The Lifeline as a Research Technique

Each participant was asked to engage in a one-time, in-depth, semi-structured interview that was intended to let the participants express their own thoughts, feelings, and attitudes. With my help, each participant filled out a “lifeline” (Baxter, 1999). The beginning of the lifeline was marked by the adolescent’s first recognition that something was “wrong” in the family. The end of the lifeline was marked by the present time of the interview. The participant was asked to tell her/his story of living with HD and being at-risk for HD using the lifeline as a guide. Only two specific questions were asked uniformly across all interviews. The first question came at the beginning of the interview when I asked, Can you tell me when you first learned about Huntington’s Disease? The second specific question came near the end of the interview when I asked, When you look into the future how would you describe your future self, and does your at-risk status impact this picture of future self? The adolescents were encouraged to talk about what they felt was most important and to move the conversation in any direction. I endeavoured to create a comfortable atmosphere in the interview process, and used the words that the adolescents used in shaping my questions and conversations. For instance, when an adolescent responded to my first question they spoke about knowing that “something was wrong in the family.” I would paraphrase their statements using the words “something wrong in the family” to help them describe their own story in a way that was natural to them. Mischler (1991) suggests

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15 Kerwin et al. (1993) identified the purpose of exploratory research as a way of investigating and identifying important variables, and of understanding little known phenomena.

16 These questions evolved over the course of the interviews and were not necessarily asked in the way they are presented here in each instance. The essential focus of the questions, however, was consistent.
that allowing the participants themselves to pattern the timing, sequence, content, and context of topics discussed enhances the validity of participant accounts.

My decision to use a lifeline as a research technique evolved from my work with UBC students in the early 1980s. I was participating in a practicum with the Department of Counselling Psychology at UBC that was intended to assist students in moving forward in their chosen career upon graduation. A decision-making line was utilized in the counselling process whereby students noted important decisions they had made (e.g., deciding to go to UBC) and placed the decisions on a line that represented their age at the time of the first significant decision identified and at the time of the last significant decision. This process enabled the students to use the line as a way to track their decisions. I found it helpful in my role as therapist as it gave me a guide to help the students explore past decisions and examine the “how” of making decisions over time. It was not unusual for students to discover that they had made many past decisions that proved to be good decisions. When they looked at the decision line and the number of decisions they had made in their life, they would often feel empowered to make the important career decision that was facing them in the present.

In considering how to frame the interview process with the adolescents in this research, I looked for a tool that would allow for direct interaction between the adolescents and myself, and that would help the adolescents explore the topic broadly yet with some structure (Baxter, 1999). It has been my experience as a therapist with adolescents that having some structure in a therapeutic session assists young clients in exploring challenging feelings and situations in their lives. Additionally, I knew that I wanted to explore the nature of “future self” with the adolescents who are at 50% risk for late onset disease. The lifeline would not only assist the adolescents in telling their stories but would also function to graphically place the question of the future on a line; they would see their own life story as a continuum, and might possibly find this
helpful when considering the unknown territory of their future lives. I felt that it would also assist me with analyzing and synthesizing their stories.

The lifeline became the initial guiding tool of the interviews. I would ask the participants to place their experiences on the lifeline, which would take approximately thirty minutes, and the rest of the interview was spent looking at the lifeline and explicating each of the incidents more fully. During the second part of the interview it was not unusual for the participants to remember events and add them to the lifeline. There was no formal structure used outside of the lifeline. Both the participant and myself were free to move the conversation in any direction.

Stephanie's lifeline (See Figure 1) is given as an example of how the lifeline was designed, and how the participants utilized it. The adolescents' lifelines and stories can be found in APPENDIX IV.

FIGURE 1: STEPHANIE'S LIFELINE

Stephanie: Age 14

- Knew something was wrong
- Two incidents of Dad being violent with Mom, Grandmother, and me
- Attend monthly adolescent-at-risk meetings with HD society
- Dad hospitalized for two weeks and put on medication
- Dad's still calm but I still don't trust him
- Getting more in trouble with Mom

AGE  8  9  10  11  12  13  14

- Dad had predictive testing
- Mom brings big book on HD
- Told HD is hereditary
- Mom put me on waiting list for psychiatrist
- See Dad's movements
- Incident at lake with Dad and Grandmother
- Friends notice Dad is different
- Dad lost licence

- Dad given more medication to calm down
- Told friends about Dad's HD. Now all friends know
- Attend national HD conference in Saskatoon
Stephanie’s lifeline begins at age eight when she recognized that “something was wrong.” We continued to mark events that she believed demonstrated the development of her awareness of HD and its impact on her life.

Each of the interviews was approximately three hours long. Upon completion of the interview the participants reviewed their own narrative accounts for accuracy. I gave the adolescents my telephone number and my e-mail address so that they could contact me if they wanted to add something to the interview, or if any feelings of discomfort resulted from the interview experience and they felt the need to talk. Six of the adolescents contacted me, and I phoned, e-mailed, or met with them. Those who contacted me were concerned about family conflict or wanted to discuss the pros and cons of predictive testing.

Fieldwork and Other Sources of Data

As mentioned previously, I was involved in a two-year internship in the Department of Medical Genetics Huntington’s Disease Clinic and Predictive Testing Centre at the University of British Columbia. Typically I would arrive at the clinic at 7:30 a.m. and leave at 6:30 p.m. I did this one day per week for two years. While at the clinic I was engaged primarily in therapy sessions with predictive test candidates, individuals at-risk for HD, HD families, at-risk children, and support people in the lives of individuals or families struggling with issues surrounding HD. I learned about the challenges of living with Huntington’s Disease from my clients. These challenges included dealing with the psychological symptoms of HD, such as mood swings and difficulty in controlling anger, and with the difficulty of distinguishing between HD symptoms and other behavioural issues. Other important dilemmas addressed included how to communicate with symptomatic family members, how to talk about the hereditary nature of HD to children, decision-making around choosing to have or not have predictive testing, to have or not have children, and a myriad of other life issues.
The clients at the clinic became my teachers. It was through them that I began to learn about how HD impacts families, and I felt touched by their trust and their ability to confront their demons. My role as a therapist in the HD community gave me a depth of understanding that I believe influenced my research. The experience helped me to focus my enquiry; I began to see what the questions were, what the important issues were.

Before and during the research, I was involved as a volunteer staff person with the British Columbia Huntington's Retreat. The six-day retreat is organized by Susan Tolley, Director of the HD Resource Centre, and has been operating for fifteen years. The purpose of the camp is to provide adults who are symptomatic with recreational and therapeutic activities, as well as a break from the stresses of everyday life. It also provides a respite for the families of the retreat participants. Many of the families use the time as an opportunity to take a vacation or time off work to renew their energy.

There are usually about fifteen people attending the retreat. They are generally mid-stage with Huntington’s Disease. Most of the volunteers have been involved for a number of years, and many of the retreat participants have attended for three or four years. The retreat provides a time when volunteers and participants come together as a community that reflects the nature of an extended family (Cox, 1999). The experience of living with retreat participants twenty-four hours a day helped me understand some of the difficulties that family members face daily, such as how to structure conversation when a participant begins to perseverate,17 or how to assist a participant when choking results from eating certain types of foods.

The intimacy inherent in the retreat experience again allowed me to enter the HD community in a way that is almost impossible for me to describe. As stated earlier, the duality of my role as researcher/therapist presented challenges, and the retreat environment was no

17 A person with HD may perseverate – become “stuck” on one idea or activity. Individuals may become rigid in their behaviour and unable to change easily from one activity or idea to another, or to alter their routine (Paulsen, 1999).
exception. As a therapist, I found myself in a situation where I was living with my clients for a week and assisting them in daily functioning. This experience gave me invaluable insight into the courage and capacity of individuals who are challenged by Huntington’s Disease. Consequently, I was aware throughout the interview process that the questions that arose for me from use of the lifeline and from listening to the adolescents’ stories were influenced by my own history within the HD community.

**Process of Analysis**

**Framing of the Interview Question**

The overall methodology of this study and the framing of the research problem was consistent with general phenomenological principles as articulated by Van Manen (1984). A modified narrative approach was also adopted to help structure the main interview questions and to inform aspects of the data analysis; that is, the main interview questions, which were derived from the general research problem (i.e., What is the experience of at-risk adolescents in a family with HD?), were framed from Cochran and Claspell’s (1987) dramaturgical-phenomenological approach to narrative research. In this approach, the researcher uses the initial data collection interview as an opportunity to elicit participants’ recollections of the investigated phenomenon as if their experience were a story with descriptive detail and temporal development. Also, during the data analysis, the fundamental structure of the participants' experiences (i.e., the essential structure of their constructions or interpretations of their experiences of living at risk for HD and in a family with HD over the course of their lives) was conceptualized in basic narrative form (i.e., as a story). The fundamental structure or common story of their experiences is presented in Chapter IV with Marilyn’s story.
Phenomenological Analysis

Phenomenological analysis involves two major steps: conducting thematic analysis, and determining essential themes. A theme is defined by Van Manen as a description of the structure of the lived experience: "themes are more like knots in the webs of our experiences, around which certain lived experiences are spun and thus experienced as meaningful wholes" (1984:20).

The process of uncovering themes in the descriptions required reviewing the data carefully and repeatedly. One challenge of phenomenological research is that it generates a large quantity of interview notes, tape recordings, jottings, or other records, all of which are a part of the analysis. Analysis is also necessarily messy, as "data" does not tend to fall into neat categories, and there can be many ways of linking different parts of discussions or observations. Analysis of the themes began with each interview, and continued across all the interviews and the reading and rereading of the transcripts.

Interview material and observations were converted to text by transcription. I transcribed the first interview myself so that I could understand the process of transcription before hiring a transcriber. I duplicated the transcription tapes to ensure that I would not lose the data. Additionally, I reviewed randomly selected portions of each of the transcripts against the tapes in order to check for accuracy, completeness, and words or phrases that the transcriber was unable to decipher because of background noises or inability to understand the adolescents' phrases.

A narrative perspective was included in the general phenomenological design for this study because of the utility of that perspective in studying experiences across the life span. The inclusion of a narrative approach is especially appropriate for research focused on individual life experiences, given that we live and make sense of our lives as a series of discrete stories, each with a beginning, a middle, and an end (Cochran, 1990; Cochran & Claspell, 1987; McAdams, 1988; Mair, 1988; Polkinghorne, 1988). The "end" does not mean that the participant's life or
growth is over, but that there is a closure inherent in the individual’s understanding of how the present has been shaped by the past. In Cohler’s (1982) view, “lives are organized in the same manner as other narratives . . . and are understandable according to the same socially shared definitions of a sensible or followable presentation” (1982:207).

Indeed, as a number of theorists (e.g., Howard, 1989, 1991; Mair, 1988; McAdams, 1988; Polkinghorne, 1988; Sarbin, 1986) have proposed, identity development itself is a process of life-story construction. Furthermore, the importance of such a story lies in its revelation of “how change from beginning to ‘end’ takes place” (Freeman, 1984:9). According to Freeman, “more than a simple mapping of discrete and isolated events – whether they be particular or general– [the study of the life course] is, in a distinct sense, an ongoing story to be told” (1984:3).

Agar (1980) discusses the problems inherent in the analysis of life history narrative, noting the tensions between ethnography and theory. Life history interviews require the researcher to understand the entire interview from “the situation that produced it, the person who gave it, and the shared and idiosyncratic knowledge which constitutes the unseen context needed to interpret it” (Agar, 1980:224). In an attempt to do justice to the analysis of these lifelines each transcribed interview was read completely to get a sense of the whole. The interview was reread and divided into units denoted by a change in subject matter or a change in activities being described. These changes were compared to the events that were placed on the lifeline as an additional aid to understanding the changes that occurred in the transcribed interview. Individual units from each interview were coded using topical codes that arose from the transcripts. The process of coding continued until all data were classified. The codes were grouped into clusters of similar topics and recoded using interpretive codes. The interpretive codes were grouped to reflect themes or patterns that emerged from the data.

As the themes emerged, I constructed a narrative life history that was compiled largely from the words of the adolescent. The stories were written to capture in more
phenomenologically sensitive terms ideas from the research process. As the themes began to emerge from the analysis they were clustered and grouped into three categories:

1. Naming the Legacy: Understanding and Misunderstanding
2. Experiencing the Legacy: Huntington’s Disease in Relation to Relationships
3. Integrating the Legacy: At the Crossroads of Self and Future Self

The advantages of using a thematic perspective were threefold: First, it facilitated the use of quotes and examples relating to particular aspects of each theme. Second, the focus on the three themes made it easier to develop a theoretical discussion of the material and link it to previous research. Third, concentrating on discrete themes within the material enabled me to meet one of the aims of the research: to offer parents and professionals specific and applicable knowledge.

Limitations of the Study

As LeCompte and Goetz (1982) observed, “attaining absolute validity and reliability is an impossible goal for any research model!” (1982:55). Nonetheless, phenomenological and other qualitative researchers approach these objectives through the conscientious use of strategies designed to enhance the credibility of their studies within the context of their particular research problems and goals (LeCompte and Goetz, 1982). In this study, attention was paid to a variety of factors that may have threatened the essential trustworthiness of the research. At the same time, certain limitations do exist on the reliability and validity of results, and, therefore, on their generalizability to the larger population of adolescents at risk for HD and other conditions.

Guba and Lincoln (1994) proposed four criteria for judging the soundness of qualitative research: credibility, transferability, dependability, and confirmability. The criterion for credibility involves establishing that the results of the research are credible from the perspective of the participant in the research. Since the qualitative perspective is to describe or understand the phenomena of interest from the participant’s viewpoint, the participants are the only ones
who can legitimately verify the credibility of the results. In this research, each participant was asked to review and comment on the summary of the interview and the summary of the story (life history) that I compiled from their interview. All of the adolescents in this research concurred with the credibility of the summary of their life history.

A second soundness criterion examines the transferability of the data. How applicable are the findings to cases outside of the research scope? This term replaces the traditional construct of external validity, which examines the generalizability of the research in quantitative studies. Often transferability/generalizability is problematic in qualitative studies due to the subjective nature of the human experience that is the integral part of the research. Additionally, in this research, only one male was interviewed. Gender is a salient variable in the socialization process, affecting how people think of themselves and how others respond to them (Gilligan, 1982; Katz, 1990; Bem, 1993). While there exists a potential for bias in the research associated with having eight females out of nine participants and recruiting all the participants from the Huntington society, it is suggested that by keeping methods transparent through the use of records, and a strong theoretical framework, other researchers can determine for themselves whether or not generalizations across to their own research are possible. Phenomenological research method is not about repeatability of the results but rather about the process of tracing how understanding unfolds. Based on the observation that data from different sources helps to corroborate, elaborate, and illuminate research, triangulation of data sources was used to enhance transferability (Marshall & Rossman, 1995).

Dependability is the third criterion. Does the research account for changing conditions in the study topic as well as changes in the research design created by a refined understanding of the setting? Marshall and Rossman emphasize that the concept of dependability is very different from that of reliability: "the positivist notions of reliability assume an unchanging universe where inquiry could, quite logically, be replicated. This assumption of an unchanging world is in
direct contrast to the qualitative/interpretative assumption that the social world is always being constructed, and the concept of replication itself is problematic” (1995:145).

Qualitative research assumes that each researcher brings a unique perspective to the research. Confirmability refers to the degree to which the results can be confirmed or corroborated by others. While similar to the positivist requirement of objectivity confirmability emphasizes the integrity of the data itself, rather than requiring the researcher to be unrealistically objective in their interpretation. Recognizing that qualitative research will always be shaped by the subjectivity of the researcher, confirmability asks the data help to confirm the general findings and lead to the stated implications (Marshall & Rossman, 1995).

There are a number of strategies for enhancing confirmability. In this research, I actively searched for and described negative instances that contradicted prior observations. Additionally, I conducted a data audit that examined the data collection and analysis procedures. As described above, I reviewed randomly selected transcripts against the tapes in order to check for accuracy and completeness. Ultimately, as Lauterbach (1992:156) states, “the real tests of reliability and validity are in the hands of readers who have experience of the phenomenon either through personal or professional experience.”

Summary

The purpose of this study is to understand the experience of adolescents at risk for Huntington’s Disease. As indicated in Chapters I and II, previous investigations on adolescents in HD families have neglected to investigate the experience of adolescents at risk for HD. Instead, investigations have been conducted in a manner where most of the information pertaining to adolescents was anecdotal and from sources other than the adolescents themselves. The phenomenological method was chosen for this research because phenomenology aims at
describing and understanding phenomena (e.g. events, occurrences, experiences) as they are lived and experienced individuals (Van Manen, 1984).

Throughout this research I experienced a dual role as researcher/therapist. Gottlieb’s (1994) decision-making model assisted me in setting appropriate boundaries while interviewing the participants in this research. My fieldwork experience of working with HD families influenced my interview questions and becomes relevant in later discussions on the implications of this research for parents and professionals.

Because my participants were adolescents it necessitated inclusion of parental consent and careful attention to informed consent and creating and ensuring confidentiality for the adolescents. The lifeline was utilized as an organizing tool to assist the adolescents in telling their story and to help them to visualize the future. The interviews were converted to text by transcription, and the summaries of the interviews were reviewed and validated by the adolescents. Although the overall methodology and the framing of the research questions were consistent with general phenomenological principles, a narrative approach was also adopted to help frame the emerging descriptions of the experiences.

The themes that emerged from the analysis were clustered and grouped into three categories: (1) Naming the Legacy: Understanding and Misunderstanding, (2) Experiencing the Legacy: Huntington’s Disease in Relation to Relationships, (3) Integrating the Legacy: At the Crossroads of Self and Future Self.

Chapter IV will utilize Marilyn’s story to illuminate the general themes that emerged from all of the stories. Through her story the reader is brought closer to the lived experience of adolescents at risk for HD.
CHAPTER IV
LIFELINES AND STORIES

The thoughts of youth are long, long thoughts.

Henry Wadsworth Longfellow

Stories give our lives form and voice. As Cochran says, “We live in story whether we like it or not. We experience life as a narrative flow” (1986:3). The stories in this research exemplify the variation and complexity of the adolescent experience of Huntington’s Disease in the family.

Purpose and Outline of Chapter

This chapter discusses the lifelines and stories that emerged from the interview process. Marilyn’s narrative will be used as an example of the stories the adolescents told about their experience of being at-risk adolescents living in a family with Huntington’s Disease. Marilyn’s story was chosen because it reflects the themes that emerged across all the adolescents’ stories, and also because it assists in identifying the individual “textural portrayal” of those themes (Marshall & Rossman, 1995). Marilyn’s story will illustrate and present the three themes that emerged from the stories and that characterize the experiences of adolescents in families with HD: (1) Naming the Legacy: Understanding and Misunderstanding; (2) Experiencing the Legacy: Huntington’s Disease in Relation to Relationships; and (3) Integrating the Legacy: At the Crossroads of Self and Future Self

Researcher as Storyteller

I see my role in this research as a storyteller or, more precisely, as a gatherer of stories. Polkinghorne (1988, 1995) argues that qualitative research involves sensitivity to stories. Inherent in that sensitivity is the understanding that this research is a story about stories. My role
as storyteller is to re-tell the stories that I have been told. The idea is not to be precise in the re-telling but rather to elucidate the essence of the at-risk adolescent’s experience of living in a family with Huntington’s Disease.

The stories emerged through the structure of the lifeline, which provided a guide and direction of sorts – a way for the conversation to begin to travel across the memories of the adolescents’ experience of living in a family with HD. Rorty (1980) argues that in the social sciences we do not hold a “mirror to nature,” rather we participate in a conversation. I became aware that neither the adolescents nor myself led the conversation; the conversations had a life of their own. As Gadamer (1975:383) points out, “a conversation has a spirit of its own, and the language in which it is conducted bears its own truth within it – i.e., that it allows something to ‘emerge’ which henceforth exists.”

The lifeline became the instrument through which the adolescents’ narrative accounts began to reflect themes characterizing the experiences of at-risk adolescents living in an HD family. The narrative accounts were based in telling stories. The stories followed the lifeline from the earliest memories of HD, or of knowing that something was “wrong,” to the present. McLeod (1997) describes how stories organize and convey our experience of the world in a way that integrates action over time. The lifeline provided the organization – the instrument – to convey experience over time. It became a frame whereby the story of the stories could begin to emerge.

**Marilyn’s Lifeline and Story**

Qualitative research seeks to extract common or general categories across all participants. I have chosen to outline Marilyn’s lifeline and story as it contains common experiences and
dynamics that recurred in all the stories of the adolescents interviewed. I believe that Marilyn's story captures the "essence" of the at-risk adolescent's experience of living in a family with Huntington's Disease. Her story characterizes the themes of "knowing something is wrong," which includes learning about HD both factually and experientially; the influence of family secrets and misunderstandings; the impact of HD on relationships both familial and peer; and the impact of HD on self-understanding and future self.

Although Marilyn's story is used to illustrate the at-risk adolescent experience of living in a family with HD, individual differences exist within each of the three themes outlined. Group characteristics evolved based on the shared dynamic of HD; however, the way such general characteristics manifested is most accurately reflected in the individual stories. For instance, every adolescent told stories of how HD impacted both familial and peer relationships. What was more complex was the depiction of the way in which HD affected the relationships in their lives.

Salience of the Adolescents' Stories

Given that Huntington's Disease is an inherited condition, and given the severity of its multidimensional symptoms, it could be said that the challenges facing a family with Huntington's Disease are probably unmatched by those of any other chronic illness (e.g., Multiple Sclerosis). The interplay of these elements contributed to the conversations and stories that emerged in the interview process. Recording the individual stories of adolescents in HD families, as well as their more widely shared collective experiences, appears then to be salient.

Marilyn's family lives near the community where I work and where my practice is based. For a variety of reasons I had a great deal of prior knowledge about the family and their concerns for Marilyn. I spoke to Marilyn and her parents about these concerns. Marilyn wanted to tell her

18 Marilyn was the third adolescent interviewed for this study. Her name and specific identifying descriptions of her life have been changed to protect her anonymity.
story to me as long as she could be assured she would be able to speak to me alone. We arranged for the interview to be conducted at my office outside of office hours. The stories and lifelines of each of the participants are in Appendix IV.

FIGURE 2. MARILYN’S LIFELINE

Marilyn: Age 16

- Is told Grandfather is sick in hospital – he was scary
- Aunt was moving funny
- Started listening a lot more to family talk
- Aunt lost job and had accidents with car
- HD still just a name
- Friends thought Dad was gay
- Embarrassed
- Associated Dad’s behaviour with aunt’s movements
- Video of Dad climbing temple – saw HD movements

AGE 7 8 9 10 11 12 13 14 15 16

- Summer vacation, Dad depressed, I figure out he has HD
- Told brother I thought Dad had HD
- Aunt told me Dad had HD – she thought family had told me
- Told about HD and its hereditary nature – day before 16th birthday
- Brother learned of HD
- Grandmother would not believe Dad had HD
- Everybody feeling sorry for me
- Dad quits work
- Everything changed – I really hate him

Marilyn’s Story

My whole life I knew it but I didn’t want to think it and now I know it. I know it is true and that is hard to deal with.

Marilyn recalls two vivid memories from her childhood. In one, her grandfather is in a room with a smell that is “sickness and dying.” The other involves her aunt’s attendance at Sunday night dinners, where she sat next to her and “ate funny and moved funny.” Her memory of her grandfather was that he was “scary, he looked like a monster.” She thought her aunt’s
behaviour was “just dumb.” One of her concerns was that her grandfather’s sickness was contagious. She did not talk about that with anyone.

When Marilyn was ten her grandfather died, at which point she was told that he had Huntington’s Disease. That same year she found out that her aunt also had HD.

I kind of clicked the two together. I thought, well, no. Because every time I kind of clicked two things, I would deny it.

Marilyn described her family as “secretive.” She would not talk to anyone because it made her feel “kind of uncomfortable talking about it in front of them because we’re so secretive.” The one person that Marilyn felt comfortable talking to was her mother. “She is pretty much the only one in the family that really isn’t that way.”

When Marilyn was twelve she became aware of “a lot more things.” She entered high school and was starting to make new friends. “They kind of thought my dad was weird.” She says that now when people meet her dad they say, “Why does he have such a high voice and it’s just, like, shut up – I’m sick of it.” Marilyn says that she tells them that he is dying of a disease. “That shuts them up.” She feels it is better to let them know that he has a disease he is dying from than to have them think he is “gay.”

Marilyn did not know if her father had taken predictive testing but assumes he did because he “found out in 1996.” Although Marilyn had been given information, she was confused about whether she was at 25% risk or 50% risk, and found the interview helpful in resolving her confusion by the information that was provided in the interview.\(^9\) It was at Christmas, when Marilyn was twelve, that she “put everything together.” She remembers her aunt almost in tears after the family talked with her. “I thought something’s up. I asked my mom.

\(^9\) None of the adolescents in the study had all the facts about HD correct, and therefore found the interview helpful in clarifying information. This point will be addressed in Chapter V.
She told me. She’s the only one that tells me anything.” Marilyn feels that she has to figure out what the question is before she can get an answer: “It’s kind of like I’m a detective.”

In the spring of Marilyn’s twelfth year the family traveled to South America, where they visited a famous temple. Marilyn and her father climbed the stairs to the top of the temple while her mother captured the trek on a video camera. When the family returned home her father watched the video.

That’s when he kind of saw himself, you know, with it. And he thought, Oh, my God. I might have been tired but I couldn’t have been that tired. So, that’s why they got testing.

In the summer of 1996 the family vacationed at a remote resort. Marilyn was thirteen years old, and she noticed that her father was behaving differently. “He would just sit there, you know, and look like he was about to die. He would not talk.” She realized her dad was depressed, and tried to figure out what the problem was. Her mother told her that he was “going through a really hard time.”

I thought, a hard time? What is he going through? And I was kind of trying to picture it. Was it work? No, it was not work. I don’t know why I figured it out but I just guessed. And then I thought, that’s what it is! That’s it!

She told her younger brother but he disagreed with her. When asked why she didn’t ask her mother she replied, “I guess I really didn’t want to know.”

When Marilyn was fifteen years old she realized that her friends thought that there was something wrong with her father. (“Why does your Dad have such a high pitched voice? He sounds like your brother. He walks funny.”) Marilyn felt that they “didn’t mean anything by it.” She became increasingly uncomfortable with being with her father in public: “Like, if he comes for parent-teacher interviews with my mom, it’s like, Oh God – Don’t!” She explained that she did not want attention drawn to her by her father’s conduct. She feels that he has always been
“kind of annoying but now with HD he’s really annoying.” Her way of coping with her father’s behaviour is to “avoid contact.”

I try to avoid any contact with other people and him. That’s why I never bring friends over anymore. It’s just too awkward because he asks them all these questions. He’ll ask them the same question like five times!

As Marilyn neared her sixteenth birthday she noticed that her aunts “all of a sudden started to show interest in me. . . .not interest but, Oh, how are you?” Her aunts had never spent a great deal of time with her but now she was invited to go shopping for “anything” she wanted. When Marilyn finally asked one of her aunts about all the attention, the reply was, “I am so sorry about your Dad.” Marilyn was shocked. Although she had figured it out when she was thirteen years old, she had denied it. Marilyn reports that her response led her aunts to realize that her parents had not told her that her father had HD.

Shortly after that encounter, Marilyn spent a day shopping with her mother. “I was pretty mad all day. I wasn’t pissed off with her but with the fact that it might be true.” After she returned home her mother asked her what was wrong. Marilyn blurted out, “You know, Daddy has Huntington’s!” Her mother started crying. Her mother believed that Marilyn did not realize that her father had HD: “Oh, no. Is that what you thought? Oh, Marilyn, I’m so sorry . . .” Her mother regretted having to acknowledge to Marilyn that her father had the gene for HD. Marilyn’s response was, “No. You’re supposed to say, No, he doesn’t have it!”

Marilyn spoke about how her parents began crying, yet she had no tears. “I didn’t know how to react to it. My whole life I knew it but I didn’t want to think it and now I knew it. I knew it was true and that was hard to deal with.” Marilyn did not know what to say to her parents.

I didn’t know what to say. It was really so awkward. I was just like, ah . . . because you always think it won’t be true. And when you find out it is true, it’s almost like you’re mad not sad. I was really mad at myself for not thinking that he had it. If I had only not asked. Now that I have the evidence, knowing that it was true, like, I don’t want it.
The events with her aunt and parents occurred a few days before Marilyn’s sixteenth birthday. It wasn’t until several days later that Marilyn talked with her mother and father. When Marilyn questioned her parents about why they had kept this secret her mother replied, “We wanted you to get through your life without knowing.” Marilyn could not believe that her parents thought she was “stupid” and wouldn’t figure it out. It was at this time that the hereditary nature of HD was explained in more detail. “When I heard that you have a 50:50 chance, what I heard was it’s either Thomas or me.” After it was explained further Marilyn asked, “You mean we can both get it! But you mean we can both not get it too?” Marilyn had never thought about the inheritability of HD in those terms.

Marilyn believes that another one of her aunts has HD.
I’m pretty sure she has it. It doesn’t affect me as much now. It’s kind of like, ok, it’s another one. You know three out of five people have it. And I know she has it . . . just because I know.

Marilyn talked about her father since he quit his job.

He’s at home pretty much every single minute I’m there. It’s pretty hard because I’ve never really liked him . . . I’ve just never had a good relationship with my dad. So, now he’s changed so much that it is almost like living with another man.

She talked about how he does not help her mother around the house because he feels he “can’t do it.” She believes that it contributes to her dislike of her father because she thinks he “uses the disease to his advantage.” Marilyn fights a great deal with her father, and she thinks her mother is “frustrated.”

She’s mad at me. She says I push my dad over the top. In a way I guess I do. It’s my therapy for proving, see, that he’s a monster. I don’t mean to, but in a way that’s how I prove that he is not normal.

Marilyn believes that her father still has a choice in his behaviour, and when she pushes him she can see the disease. She also realizes that he can’t “put the effort into being the father that we need.” When asked about the last time she was with her father “without the disease” she
spoke of a time when she was eleven years old and he asked her to go roller blading in Stanley Park.

Marilyn thinks that there might be a cure by the time she becomes symptomatic. At this point in her life she is ambivalent about predictive testing.

If I do get it, I don’t really want it. Like, to live to that stage where I am in a nursing home drooling out of the side of my mouth, getting my Jello . . . I would rather live my life to the fullest and then die. I guess I associate the end with my grandfather because that’s what I saw.

Marilyn has seen that Huntington’s Disease can change peoples’ lives, and, consequently, she is going to live her life more fully.

If I establish my life now then it won’t really matter because I’ll have everything that I have dreamed and strived for before it ends. . . . Don’t have any regrets is basically how I see my future. Live my life the way I want to now.

Emergent Themes

Themes emerged from Marilyn’s story that were consistent within all the adolescents’ stories; they will be illuminated in this section but will be developed in more detail in Chapters V, VI, and VII.

Marilyn’s story begins with an early memory of her grandfather dying. Marilyn had a concern about the illness being contagious yet she never approached an adult to get clarification or reassurance. When the illness was given a label, and she became aware that her aunt had the same disease as her grandfather, she realized that there was a connection. She recognized the connection and “denied” it. She did not get clarification from the family as she saw her family as “secretive.” She learned not to ask or talk openly about the disease called “Huntington’s.”

Marilyn’s struggle with an awareness that something was wrong in the family, and her decision to keep this awareness to herself, reflects the process that many of the adolescents in this research described between the ages of five and twelve.
Knowing that “something’s wrong” and not discussing this feeling with the family is an experience that cuts across the lives of six of the nine adolescents. It reflects the first theme that emerged from all of the stories: Naming the Legacy.

All of the stories involved the aspect of knowing intuitively, being told about HD, experiencing the symptoms of a parent and/or family member, understanding as well as misunderstanding the nature of the disease and the hereditary aspect of the disease, and being involved in a family where secrets affected not only knowledge but communication within the family and the adolescent’s social world.

As Marilyn’s story continues, the complexity of relationships in the family and outside the family becomes part of the illness experience. As was the case with all the adolescents, learning to handle relationships in the family was a challenge. Whether this was entirely due to Huntington’s Disease or whether it was a reflection of the adolescent’s developmental state is explored in the next theme that emerged from the stories: Experiencing the Legacy.

The familial and non-familial relationships of the adolescents reflect the complexities of negotiating relationships with the additional stress of living with HD and at risk for HD. All of the adolescents told the story of being at risk and of dealing with the challenges of having HD present in the family through the exploration of relationships with parents (both symptomatic and non-symptomatic), siblings, extended family members, and peers.

Marilyn’s story, like the other adolescents’ stories, reflects the process of identity development – an aspect of adolescence that begins with the process of self-understanding. This is a time when physical, cognitive, and social development is such that adolescents can begin to synthesize childhood identities and build a self-concept that incorporates future possible selves.²⁰

²⁰The possible self is what individuals might become, what they would like to become, and what they are afraid of becoming (Markus & Nurius, 1986). Thus, adolescents’ possible selves include both what adolescents hope to be as well as what they dread they will become. In this view, the presence of both hoped-for as well as dreaded selves is psychologically healthy, providing a balance between positive, expected selves and negative, feared selves.
The theme “Integrating the Legacy” emerges from the stories and helps establish how experiences in family and social life contribute to self-understanding, understanding of future self, and the role HD plays in these processes.

Summary

Marilyn’s lifeline was presented in order to show how the researcher and participant engaged in the interview process, and to demonstrate the emergence of the three themes. These themes will be discussed in the next three chapters of the research and will provide an in-depth, retrospective view of the experience of the at-risk adolescent living in a family with Huntington’s Disease.
CHAPTER V

NAMING THE LEGACY: UNDERSTANDING AND MISUNDERSTANDING

HUNTINGTON’S DISEASE

I knew that something was wrong. My dad was different.

Purpose and Outline of Chapter

As Marilyn’s story demonstrated in Chapter IV, three themes emerged from the adolescent’s stories, “Naming the Legacy,” “Experiencing the Legacy,” and “Integrating the Legacy.” In this chapter the adolescents’ process of moving from knowing that “something’s wrong” to integrating the practical knowledge and experience of Huntington’s Disease in their lives will be detailed. There are three stages in this learning process: (1) intuition, (2) practical and experiential learning, and (3) understanding and misunderstanding.

The chapter begins with an exploration of the adolescents’ intuition that “something is wrong.” This realization was usually the result of perceived changes in the behaviour of a parent or relative (i.e., the manifestation of symptoms) or in the family dynamic, or because access to a parent was cut off. The next section of the chapter will explore who told the adolescents about HD, and illuminate the second stage of learning about the disease. At this stage, all of the adolescents demonstrated two types of learning: practical learning and experiential learning. The practical learning involved being told by an adult that a parent had a disease called Huntington’s; the experiential learning came when the adolescents began to experience their parent’s symptoms of HD and to distinguish those symptoms from other behaviour.
The last section of the chapter will explore the third stage of learning about HD, which involves what the adolescents understood about HD, what they told their friends, and how they experienced family secrets and misunderstandings.

**Intuition: Sensing something is wrong**

As stated in the previous chapter, the adolescents were directed to draw a lifeline that began when they first realized that something “different,” “wrong,” or “unusual” was happening in the family. I decided on this approach based on my fieldwork where I heard stories from HD-affected families about the beginning of an awareness of HD. As the interviews progressed, I began to realize that many of the adolescents remembered feeling that something was wrong long before they were informed of HD or of the fact that HD was a hereditary disease.

Nancy Wexler (1991) has reflected on a time when she was an older teen and was aware that something was changing in her mother’s behaviour. The recognition of this change happened before she had any knowledge of HD.

> When I was little, I adored my mother as someone who gave me unquestioning love and warmth. But as I got older, I knew that something was vitally wrong. She was sad, silent, listless, and vague. It was as if some dark subterranean river was taking her away from me (1991:13).

As summarized in Table II, six of the nine adolescents initially had a sense there was something wrong. They reported their first awareness from the early age of five to the age of twelve, which is much earlier than the time period cited by Nancy Wexler. Yet, much like Wexler, these six adolescents’ awareness of something going on in the family was not associated with HD or any medical problem. Something was wrong either with the behaviour of a parent or relative or the family, or they had been denied access to a parent.
### TABLE II

**INTUITING AND LEARNING ABOUT HD AS REPORTED BY PARTICIPANTS**

<table>
<thead>
<tr>
<th>Intuiting</th>
<th>Learning about HD</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Affected Parent</strong></td>
<td><strong>Behaviour of Parent</strong></td>
</tr>
<tr>
<td>Stephanie</td>
<td>Dad</td>
</tr>
<tr>
<td>Carla</td>
<td>Mom</td>
</tr>
<tr>
<td>Marilyn</td>
<td>Dad</td>
</tr>
<tr>
<td><strong>John</strong></td>
<td>Dad</td>
</tr>
<tr>
<td><strong>Daphne</strong></td>
<td>Dad</td>
</tr>
<tr>
<td>Emma</td>
<td>Dad</td>
</tr>
<tr>
<td><strong>Tessa</strong></td>
<td>Mom</td>
</tr>
<tr>
<td><strong>Margaret</strong></td>
<td>Mom</td>
</tr>
<tr>
<td><strong>Kathy</strong></td>
<td>Mom</td>
</tr>
</tbody>
</table>

**Behaviour of parent and/or relative**

Three adolescents knew there was “something wrong” based on the behaviour of a relative or a parent. Stephanie was eight when she began to realize that her father acted differently than other fathers. She never questioned her mother about her father’s behaviour but thought that must be “the way it is.” She remembered asking herself why. It was a question she did not ask her mother. It was a question that she could not answer for herself.

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21 Age of child when event occurred is in brackets “()”. *Italics and Bold* indicate the subjects are siblings, i.e., John and Daphne are siblings.
The first time that Margaret realized that something was different in her family was at her seventh birthday party. “We had just moved into town. Nobody was aware of my mom’s disease. She had fallen down the stairs, and two of the mothers were present and took their daughters home because they thought she was drunk.” When asked how she knew her friends’ mothers thought her mom was drunk she replied, “One of them told me at school the next day. You know how little kids are. They don’t keep anything, they splat it all.” At that time Margaret “didn’t really know what was going on and why mom had fallen down the stairs but I knew she wasn’t drunk. I knew that much.”

As stated in the last chapter, two events occurred that created a concern for Marilyn. The first memory, at age seven, was of her symptomatic grandfather in the hospital. She was told he had Huntington’s Disease. Her concern was that the sickness was contagious, although she did not talk about that with anyone. The next recognition of something wrong evolved during Sunday night dinners when she noticed that her aunt “ate funny and moved funny.” She did not think of her aunt as having symptoms; she thought her behaviour was “dumb.” At age ten her grandfather died and she was told that he had died of Huntington’s Disease. During that same year she found out that her aunt had HD.

All three of the adolescents recognized that something was wrong yet did not ask for clarification from the adults in their life or their siblings. Marilyn’s story explicates the process of her confusion and the way that she coped with the information. She realized that something was happening for two members of her family. She was given the label of HD for the illness, and she then began to piece together the possibility of the hereditary nature of the disease. She felt unable to take the possibility further, and reports that she used denial as a coping method for the dissonance created by her realization. The information caused her sufficient enough distress and confusion that denial in this case became a way of coping with information she was not ready to process.
Denial of access to a parent

Daphne knew something was wrong when she was thirteen, not because she felt that her father was different from other fathers but because her mother would not let her see her father for “like, a year.” She did not know what was wrong but she did know that something was amiss and that it had to do with her father. When Daphne was fourteen she and her family met with a medical professional\(^{22}\) and she learned that her father had Huntington’s Disease. It was at that meeting that she learned HD was hereditary.

Daphne’s brother, John, was twelve when he realized that something was wrong in his family. He recounted, as did his sister, that this realization came about when he stopped seeing his father regularly and his parents were not talking to each other. Shortly after that time, John attended a meeting with a medical health professional that also included his sister, mother, father, stepmother, and his mom’s best friend. It was there that he first learned about HD, as he had no contact with any relatives that may have been symptomatic.

For both Daphne and John the inability to connect with their father became a signal that “something was wrong.” They did not relate the absence of contact with HD; it was only after the meeting with the medical health professional that the association between HD and their father’s absence became connected. They could now identify that HD was significant enough to cause family conflict and create the absence of their father in their lives.

Behaviour of the family

Carla recalled that she was five when she knew something was “wrong between our family.” She has memories of her mother yelling at her father, and of her father telling her that

\(^{22}\) Neither Daphne nor John remembered whether the medical professional was a psychiatrist, psychologist, or medical geneticist.
there was “something wrong.” She understood that the problem was a family problem because it occurred in arguments between her parents. Carla’s inability to understand what was wrong caused her to believe that “sometimes” it was her fault. It is a feeling that Carla still experiences at fifteen. She first learned about HD when she was six years old; her grandfather was in the hospital and she was told he had Huntington’s Disease. It was not until she was ten that she learned the disease was hereditary and that her mother had HD.

In summary, six adolescents felt that there was “something wrong” in their families. This realization occurred at ages ranging between five and twelve. It was not necessarily associated with HD or any medical problem: something was wrong either with the behaviour of a parent or relative or with the family as a whole, or was evidenced by the denial of access to a parent. For these six adolescents, the fact that the presence of Huntington’s Disease in the family was kept secret did not relieve them of worry. Children are intuitive, and can often sense when something is disrupting the family system. Giving this disruption a reason and a name can assist some children in coping with the challenges of HD in the family system.

**Practical and Experiential Learning: The Tellers**

“When you find out it is true, it’s almost like your mad not sad.”

As stated in the literature review, previous studies have shown that parents of young people at risk for HD feel burdened by the thought of giving genetic information to their children, and find it too great a responsibility to cope with alone (Korer & Fitzsimmons, 1985; Tyler & Harper, 1983; Barette & Marsden, 1979). As summarized in Table *1* all of the nine adolescents in this research began their practical learning when they were told about HD between the ages of nine and fourteen, with five of the adolescents being told between the ages of nine and eleven. One adolescent (Tessa) was given this information by her affected mother; two adolescents (Stephanie and Emma) were told about HD by their non-affected parent; two of three...
adolescents in the same family (Margaret and Kathy) were told by both their parents; one adolescent (Marilyn) was told by an extended family member; and three adolescents (Daphne, John, and Carla) were told by a medical health professional in the presence of other family members.

**Teller as affected parent**

Tessa was the only participant whose affected parent told her about HD and that she had the disease, unlike Tessa’s sisters, Margaret and Kathy, who were told by both parents. Tessa began her interview by talking about the time when she was ten and her mother told her that she had had HD since Tessa was three years old. Although she claimed that she was comfortable asking her mother about HD she commented that she “never talks about it.”

As the interview continued it became clear that when Tessa turned ten her life began to change. It was shortly after her mother informed her of her HD status that Tessa started to realize that her mom was “different than everyone else.” “I never really started to realize it until a number of years ago. I don’t know... when I walk places with her and stuff. She didn’t have a walker back then.”

For Tessa, as well as the other adolescents in this research, there were two parts to learning about HD. The first part came with her mother’s disclosure, which was when she was given factual information about HD. The second part was the experiential knowledge of HD that came with the understanding of how the disease manifested in everyday experience and made her mother “different.”

The two components of this learning process did not occur at the same time. The first part – the practical learning – occurred as a result of being given information from an adult. The second part – the experiential learning – was realized as the adolescents began to experience HD as symptoms manifested in their parent, and to distinguish the symptoms from other behaviour.
They may or may not be correct in their assessment of what behaviour is a symptom and what behaviour is not a symptom; however, most of the adolescents cited times when other members in the family would agree with their recognition of symptoms in the affected parent.

Although Tessa does not remember her mother when she was unaffected by HD it was only once she was told and understood that her mother had HD that she began associating the symptoms with the disease. Information about the disease allowed her to discriminate between her mother’s behaviour and the behaviour of other mothers. For Tessa, the division between learning and knowing about HD may well have been because she did not have any memories of her mother when she was not symptomatic. Interestingly, all the adolescents follow the same pattern of learning and knowing even though individual experiences differ.

**Teller as non-affected parent**

Two of the adolescents in this research were told about HD from a parent who was not affected by HD. Stephanie was eleven when her mother brought out the “big book” on HD, a large black binder that contained articles and other information on the disease. Stephanie didn’t want to hear about it. She said she wasn’t scared of the information because she was sure that a cure would be found “in, like, two years.” She saw HD as something “so far away.”

After Stephanie learned about HD her aunt came to visit, and Stephanie suspected that she had the disease. “I can tell. She’s like kind of moving, and her hands and feet are kind of going.” Her mom confirmed Stephanie’s suspicions, and that was a validation for Stephanie. She realized that what she perceived as symptoms of HD was accurate. Stephanie became party to many “family secrets” regarding HD. For instance, she knew what was put on her grandfather’s

\[23\] The information concerning the cure was given to her after her mother listened to a TV interview given by a researcher.
death notice, the status of a missing uncle, and the family's beliefs about who did and didn't have HD.

Emma learned about HD at age fourteen when her mother informed her that her father had an appointment with the doctor. "She explained to me that there was a disease that ran in the family and that my dad was going to an appointment to see if he was eligible for this disease." Emma felt that it was "no big deal." Her father did not talk about it with her, "so it was sort of a secret sort of thing." Her mother told her that it was "a disease that affects your nerves and makes you a little weird." She knew that her grandmother had died from a disease, and she believed that her aunt also had HD. "She doesn't talk about it but we all know she does. What I see in my dad, I see in her."

Emma did not see any symptoms in her father until she was fifteen and her father started "shaking." Emma began to realize that the changes she perceived in her father's behaviour were related to HD. She also spoke of a change occurring in her father's temper. She related that her father had "an unbelievable temper for years, and years, and years," and that his temper was worse now in different ways. "He used to be abusive but now he is just verbally abusive. Little things I say to him he twists around. He gets mad about them all the time. Like he is always mad. He is never happy."

Both Stephanie and Emma learned about the disease from the non-affected parent, and began to gain knowledge about the disease by identifying symptoms in their affected parent and/or other extended family members. It is interesting to note that neither Stephanie nor Emma discussed HD with their affected parent after learning about HD.

Tellers as both parents

Margaret and Kathy, who are sisters, each learned about HD at age ten from both of their parents. For Margaret the memory of being told is vague.
It’s really hard to remember because I’ve lived with it all my life. I see old videos at home with my mom – her speech is normal and her balance is normal and I don’t ever remember that part of my mom because I was so young when it happened. So, the only things I can remember are recent, like the last couple years.

At eleven Margaret’s “eyes opened” and she realized “everything.” She began to see the changes in her mother’s behaviour: “She was falling more often, her speech was slurred, and I could just tell.” She related that she talked with her parents as she witnessed the changes in order to understand and validate that the behaviours she witnessed were the symptoms of HD.

Kathy, who is older than Margaret, remembers her parents telling her about HD when she was ten years old. “I didn’t have a clue about it before. It just went in one ear and out the other.” Her parents also talked to her about the hereditary nature of HD: “I didn’t quite understand that either at the time. Hereditary – that’s a big word for a ten-year-old.” She did not see her mother as having symptoms of HD; rather, it was “just that mom was acting different.” Kathy noticed that her mother began “shaking, she stuttered and slurred and she couldn’t drive.” Kathy described “piecing it all together” as a “long process of understanding.” “I’m very visual and I just kind of watched – that’s the way I operate.” She believes that it was the same process for her sisters.24

Both sisters learned about HD at the age of ten but began to experientially know about the disease as they identified symptoms in their affected parent and experienced how it impacted their daily lives.

24 Margaret was one of three sisters involved in this research. Kathy was the oldest. Margaret was the next oldest and Tessa was the youngest. There is one older sister but she was 21 years-of-age older than the age requirements of the research.
Teller as extended family member

As described in Chapter IV, Marilyn was ten when she learned that her grandfather died of HD. During that same year she found out that her aunt had the disease. While she recognized that there was probably some significance to the fact that they both had the same disease she recalls that she would “deny it.”

Marilyn knew that her grandfather was sick, and at a young age was told he had HD. She began to perceive changes in her aunt, and at age ten was told by her mother that her aunt had HD. Marilyn deduced that there was something strange occurring with two family members having the same disease. It was the accidental disclosure by her aunt of her father’s illness that caused her to believe that the differences she perceived in her father were the result of Huntington’s Disease and that it must be hereditary.

Teller as health professional

Daphne and John knew that something was wrong when they were denied access to their father. It was not until they attended a meeting with their father and a medical health professional that they learned about HD.

John was twelve when he attended the meeting, and he described it as beginning with their father reading a letter to the children “to express his feelings.” The letter explained that he had wanted to tell them about having Huntington’s Disease but their mother had not wanted the kids to know. “My mom was pretty angry and hurt and so was everyone else. I was more like, this is about my dad having Huntington’s, and so it kind of went bad from there.”

\[\text{It is important to note that denial is not necessarily maladaptive. Bloch (1993) proposes that denial and repression can be viewed as strategies for dealing with the impact of HD. Denial allows the individual to have more time to engage in the important emotional work of slowly assimilating the information about HD that is creating a new reality in the adolescent’s life.}\]
When asked what he learned from the meeting, John commented, “I learned that he was not going to die right away . . . it usually takes a while to set on, and I have a chance of getting it.” His mother wanted to talk about the meeting but John felt he would not be ready to “for a little while.”

I don’t think it actually hit me that it could really affect me. I was just kind of worried about my dad . . . that he was going to die pretty soon. I still had the image that it was going to be a lot quicker than what they told me. So, I started talking to my mom, I think around thirteen when I actually started noticing twitching.

Both siblings characterized the meeting as going from informative to a family argument; it was clearly upsetting for both of the children. They both felt that the family argument that took place in the meeting was as difficult for them as the news they received. John felt that his parents’ fighting and conflict in the medical health professional’s office affected him more than the disclosure of HD. “I was pretty stressed out already as a kid. I wanted to kind of forget about it [HD]. There was one too many things to think about in my life.”

Daphne was fourteen at the time of the meeting and recalled that “Mom was there and Dad actually told us.” She recounted that her father’s letter was “about how it affected him – not being able to see us because of this disease, and how much it hurt him and all this kind of stuff, too, which was valid. . . . He blamed my mom and it is not her fault.”

Daphne felt that the meeting was a hard place to learn about her father’s disease. I would have rather known right up front. She’s always protected us but I have always hated it because I don’t feel I need to be protected. I would rather have had the opportunity to do it myself and to seek out help if I needed it or not.

The meeting was difficult: “It was like all this family stuff on top of it. Like no one can understand the family dynamics. We’re all crazy.” When asked what she learned about Huntington’s from the meeting she stated, “Well, it was that I have a good chance of getting it and that my dad is going to die from it.”
It is interesting to note that Daphne felt she had a “good” chance of getting the disease. A year after the interview Daphne called and asked to meet with me. We discussed her indecision about testing; she felt that she would have the gene for HD and that her brother would not. This was the first time in all of the interviews I conducted that an adolescent spoke to me about feeling that they were “preselected” for the disease. Daphne told me that she felt she was more like her father than her brother was. She felt her brother did not have the gene because he was much more like her mother.

Daphne reported in her interview that she had problems dealing with her emotions and had attempted suicide. Against her will she was placed in a psychiatric unit for a period of time. She identified strongly with her grandmother, who was diagnosed as having a manic depressive disorder, and she felt that she had much more difficulty dealing with life than the rest of her family. It seems that Daphne may have preselected herself based on her associations with family members who had HD or mental illness. I do not know if the rest of the family participated in the preselection, and, consequently, assigned her the sick role.

Both Daphne and John learned about HD in a meeting with a medical health professional. John walked away from the meeting upset about learning about HD, and, moreover, by the argument that ensued in the meeting. He felt his father would die sooner than he was told, and that he was at risk for HD. It was not until a year later, when he saw his father’s movements, that he began to experience the reality of the disease. It was at this point that he could begin to talk with his mother about his feelings.

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26 Preselection refers to the practice of deciding who is going to develop the disease in the family prior to age of onset, and is a phenomenon that occurs frequently in families when there are early signs of onset of HD (Korer & Fitzsimmons, 1985). Further, Kessler and Bloch (1989) argue that preselection delegates one family member to the “sick role” (Parsons, 1952) and therefore the remaining family members become organized around the beliefs that are consistent with that assignment.
Daphne also left the meeting upset about what she learned, as well as with the argument that occurred. She heard that her father would die, and that she had a “good” chance of having the gene for HD. She did not choose to speak to her mother about her feelings when she began to notice her father’s movements: “He was pretty shaky sometimes and just dropping things that he would not normally drop. Or his memory – his short-term memory got a lot worse and I had to remind him of things.”

Carla recounted when the family had attended a predictive testing session and she was informed about HD by the genetic counsellor. It was only afterwards that her father talked with her about HD, and the fact that it was hereditary. All she remembers is that she wanted to know if her mother was “going to die tomorrow,” and that she had no idea what to ask. “I walked away thinking that my mom was dying and I had no idea when. And I didn’t care that I had it, that I had a chance. I was just like, no, my mom can’t leave me.”

When Carla was fourteen she attended a tour at the Centre for Molecular Medicine and Therapeutics with the HD Society. Huntington’s Disease was explained in detail, and there were several question-and-answer periods. “I knew that HD was hereditary, but I learned the most about it on the tour.” It was also at this time that Carla’s mother was placed in an extended care facility. “Even though I knew it was a deadly disease, I always kind of had that hope that she’d get better, but when she went into the care home it was definitely like, that’s it – there’s no hope.”

Carla also reflects the two-tiered process of learning about Huntington’s. She had learned about the disease in the first meeting at the predictive testing clinic and in the ensuing conversation with her father. It was when her mother was placed in a care facility that she understood the reality of her mother’s illness. She was also able to ask questions at the Centre for Molecular Medicine and Therapeutics that helped her understand the significance of the hereditary nature of the disease. It may well be that because Carla had grown up believing that
something was wrong with the family, and that she was a part of what was wrong, it caused her to see her mother's symptoms as partially due to family conflict. When her mother entered the extended care facility she could no longer view the disease as only a family issue. Her mother was no longer able to function on her own, and Carla could no longer delude herself that her mother would improve. This experience enabled her to ask questions and begin to understand the hereditary nature of HD.

In summary, the nine adolescents in this study were told about Huntington's Disease between the ages of nine and fourteen. Although they learned by different means (family member or a medical professional with family present), all of the adolescents demonstrated two levels to their learning: practical learning and experiential learning. The first level involved learning from an adult that a parent had a disease called HD, and the second level of learning occurred when the adolescents began to see HD symptoms in a parent or relative and to distinguish the symptoms from other behaviour. Carla's experience was the only slight variation to this process of experiential learning, as she reported that it was her mother's move into the extended care facility that caused her to realize the extent of damage caused by HD.

**Understanding and Misunderstanding: Explanations and Secrets**

Although a family member or a medical professional told the adolescents about the presence of Huntington's Disease in the family, there was little discussion about HD beyond this first explanation. Many of the adolescents expressed that their family had "secrets," or they would describe the family as "secretive." Most of the adolescents had misunderstandings or unanswered questions about HD. In each interview I would explain, correct, or provide

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27 Additionally, some of the adolescents reported that they did not want to talk about the disease with the family. There were three explanations offered: they "didn't care," there will be a "cure," or it is "far away."
information pamphlets when a question came up or it was obvious that there was a misunderstanding about HD.

This section will explore the adolescents' understanding of HD and how this understanding is revealed to their peers. Secondly, I will explore how secrets and misunderstandings affect this knowledge. The findings are summarized in Table III.

Adolescent Understanding of Huntington's Disease

Most people at risk for HD are what Nancy Wexler (1979) calls “disease wise” – that is, from a very early age they know what the disease is called and some basic facts about its inheritance, that it is “in the blood” or “passed down in families,” for example. I explored with each adolescent in this study what he or she understood about HD, but I was also interested in how they explained HD to their friends. In my work with adolescents I have learned that what young people know and how they explain it to their peers may be quite different. It is not unusual for an adolescent client to ask for assistance with formulating what to say to their friends when there has been a death or divorce in the family. It occurred to me that it might well be the same when an adolescent tries to explain a parent’s HD symptoms to their friends.

Stephanie said her mother told her that HD was a “degenerate brain disease,” and that her father’s brain cells were “slowly dying.”

And I was kind of being like, what is that? Ok, whatever. And then we just kind of talked about it for a bit and she explained how long they figured he had it for . . . the causes of it . . . because he had an accident and that brought it on earlier.
### TABLE III
NAMING THE LEGACY SUMMARY
UNDERSTANDING AND MISUNDERSTANDINGS

<table>
<thead>
<tr>
<th>Name</th>
<th>Basic understanding</th>
<th>As teller</th>
<th>Secrets - bonding</th>
<th>Secrets and misunderstandings</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Stephanie</strong></td>
<td>Head Injury accident explained as cause of onset</td>
<td>Family folklore Factual Explains parent's symptoms</td>
<td>Bond with mom Withholds from dad and father's family</td>
<td></td>
</tr>
<tr>
<td><strong>Carla</strong></td>
<td>Factual Witnessing symptoms</td>
<td>Reacts to friends Explains symptoms to relieve social stigma - as &quot;not being dangerous&quot;</td>
<td></td>
<td>Fear of juvenile HD Did not ask parents about misunderstanding</td>
</tr>
<tr>
<td><strong>Marilyn</strong></td>
<td>Factual Witnessing symptoms</td>
<td>Reacts to friends Explains symptoms to relieve social stigma - as &quot;not being gay&quot;</td>
<td>Figured things out Bond with mom Mom is information source</td>
<td>Confused about the odds of carrying the gene Bond with mom lead to clarification</td>
</tr>
<tr>
<td><strong>John</strong></td>
<td>Factual Witnessing symptoms</td>
<td>Parkinson’s as a metaphor</td>
<td>Bond with mom when symptoms appeared. No dialogue with father</td>
<td></td>
</tr>
<tr>
<td><strong>Daphne</strong></td>
<td>Factual Witnessing symptoms HD Support group</td>
<td>Alzheimer’s as a metaphor</td>
<td>No dialogue with mother or father. Bonded with grandmother</td>
<td></td>
</tr>
<tr>
<td><strong>Emma</strong></td>
<td>Factual Witnessing symptoms</td>
<td>Reacts to friends Explains symptoms to relieve social stigma - as not being “angry or rude”</td>
<td>Figured things out Bond with mom prior to her death</td>
<td></td>
</tr>
<tr>
<td><strong>Tessa</strong></td>
<td>Factual Witnessing symptoms</td>
<td>Hesitant Factual School project</td>
<td></td>
<td>Fear of juvenile HD Did not ask parents about misunderstanding</td>
</tr>
<tr>
<td><strong>Margaret</strong></td>
<td>Factual Witnessing symptoms</td>
<td>Hesitant Factual Explains parent’s symptoms</td>
<td></td>
<td>Bond with father</td>
</tr>
<tr>
<td><strong>Kathy</strong></td>
<td>Factual Witnessing symptoms</td>
<td>Hesitant Factual Explains parent’s symptoms</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
Stephanie’s father had had a serious motorcycle accident, which her mother said was responsible for the early onset of her father’s disease. Stephanie regarded this as yet another fact about the nature of HD. It is important to note that trauma, such as that caused by a motorcycle accident, has not been demonstrated to have any effect on the onset of HD. This trauma theory could be construed as an example of the misunderstandings that surround HD, or it may be an abstracted form of knowledge that the family has used to explain why the disease has manifested itself differently with Stephanie’s father than with the rest of the family (Stephanie’s grandfather or aunt). Stephanie and her mother believe that the serious injury that resulted from the motorcycle accident was the cause of the early manifestations of HD in Stephanie’s father. Stephanie believes that, were it not for the accident, her father would not have exhibited symptoms of HD until a much later date.

The possibility that families have folklore that represents alternative forms of knowledge is not generally reflected in the literature on HD or genetic counselling. For those families with Huntington’s Disease, family folklore is built on observation and the lived experiences of the family members. During the two years that I was involved in the predictive testing program at the Huntington’s Disease Clinic at UBC Hospital I explored these forms of knowledge with HD families, and was provided a deeper understanding of the impact of living with HD.

When Stephanie was asked what she would tell her friends about HD she responded,

I would say it’s his brain cells that are slowly dying – like he is losing them abnormally, and you can transport it. There is a whole bunch of stuff on medical research that’s helping them. They get really angry and frustrated and everything kind of sets them off and it’s pretty much too late. It’s really something. Like, oh, they’re flat – really flat.
Stephanie would describe HD to her friends with more than the factual information she received from her mother. She included that it was hereditary, by using the term “transport.” Stephanie expressed some hope by describing “a whole bunch of stuff on medical research.” Lastly, she described the disease through her experiential knowledge of her father’s symptoms.

Marilyn tried to explain what HD is to her boyfriend, and it resulted in an argument around the probability that Marilyn would have the gene. Her boyfriend went so far as to talk to his biology teacher to learn how 25% risk and 50% risk is determined. Marilyn did not get upset with her boyfriend because he was trying to learn about HD. Marilyn believes that “most people just want to know about it but they don’t really understand it.”

Marilyn discussed how she decided to tell her friends about her father. Her motivation came from concern that her friends would believe that her father’s symptoms were the affectations of homosexuality.

So, when I started making new friends . . . they kind of thought my dad was weird but they never really said anything. But even now, if I meet people and they meet my dad — you know — who is Mr. __ and why does he have such a high voice, and it’s just like, shut up. I’m sick of it.

For Marilyn it is easier to deal with the fact that her father has a disease that he is dying from than to deal with the social stigma of her father being seen as “gay.”

[Loud sorrowful laugh] Oooh, I kind of just say, Oh, yeah, he’s dying because he has this disease. And they say, Oh, I’m so sorry! I don’t know what to say! That shuts them up. If it’s a certain person you kind of like, BLAM! But if there is someone you like you just kind of make it softer . . . I kind of like talking about it because I want them to know he’s not gay.

Marilyn, like Stephanie, described HD to her friends in terms beyond factual information. She described her father’s symptoms as she experienced them. She was motivated to describe his symptoms to her friends in order to avoid the stigma of homosexuality, which she perceived as

28 The adolescents in this study would often use their own terms to describe medical information.
far more detrimental than the stigma of HD. It was not only her father being perceived as a homosexual that bothered her, it was also that such a perception would reflect poorly on her.

John recalled that he was told that his father had HD, that he was not going to die, that the disease would debilitate him, and that it usually “comes really late in life but my dad has it now.” He was also told that the disease was “successive,” which was John’s term for the hereditary nature of HD.

And then it took him [the medical professional] a little while to tell me I was someone who could be like “successive,” that I could be tested. It was kind of hard to think about it – like, whoa, whoa, whoa. Like, my dad has it, so I think I asked him a question, like if I was to have kids . . . it is genetic so I could have it.

After John began to experience his father’s symptomatic behaviour he went on the Internet to find more information.

When I was fifteen or sixteen I started looking for information. I went on the Internet. I checked out all the stuff I wanted to know. I wanted to know what cures they made and all that stuff, and how much research they have done. I found out all that they have done. I found out that they had done one of the first – I can’t remember what it is called – but they found the gene. All the genetic research that is done on all kinds of other things is based on the Huntington’s model. It was one of the first or something.

The fact that research on Huntington’s Disease was creating a new protocol in predictive testing gave John some reassurance. “I found out that [the predictive testing protocol] was a good sign.” But for John, the hereditary nature of HD was what troubled him the most: “The main thing for me is that it’s a 50:50 chance. That’s the main thing.”

When asked how he explains HD to his friends, John replied,

I said it was kind of like Parkinson’s. [Laughs] People know what Parkinson’s is but a lot of people don’t know what Huntington’s is. I would explain to people by saying that Huntington’s is a genetic disease that affects some people, and that it is a debilitating disease, which means it slowly takes away things that you could normally do, like shaking your hands and starts to affect how your mood acts…how you act in everyday life, but it usually takes a while to set in and you are not for sure going to get it. But it usually does take effect when you are thirty-five to thirty-seven, and that’s a long way to wait anyway. [Laughs]
John described HD to his friends by comparing it to a disease he thought they might know. He used both factual and experiential information to describe the nature of the disease as well as the symptoms he experienced with his father. He also spent some time explaining the hereditary nature of the disease, which he stated was the “main thing” for him. When I asked him if he talked about being at risk with his friends, he replied that some of his friends had asked him if he could get the disease: “Sometimes, they ask. A couple of them asked. I said, yeah, I could get that. It was a very hard thing to say.”

Daphne (John’s sister) also described HD to her friends using another disease as an example.

Well, I describe it a lot like Alzheimer’s, the same kind or somewhat similar effects. Is it Alzheimer’s? It is gene-oriented, something having to do with the genes. I am 100% sure. And that it is hereditary, so like I can possibly get it, or my brother. It is basically movement-oriented. It is like the movements slowly get sloppy and you shake around a lot. Um, short-term memory goes first. You know you have to remind them of things constantly. Um, what else? They yell at things; he doesn’t mean to be so loud but it is because he is having trouble getting his thoughts in. That is basically how I would describe it.

Daphne used the factual information she received at her meeting with the medical professional as well as her experience of her father’s symptoms and the fact that it is a hereditary disease. She stated that she was motivated to tell her friends so that they would not be confused when they met her father.

I usually give them a little bit of warning because I know [that if] I didn’t really know what was going on I would probably be [confused]... So, I usually just give them a heads up on what’s going on because then they don’t go in and not know.

Daphne went to a meeting at the HD Society in her community. She was looking for more information on the disease, and for support from people in the same situation.

It helped me sort of identify with people who had been in the exact same situation as me. ... People who are old with it, young with it, you know? I had a lot of different people to talk to, and they are thinking of getting tested at that point.
And I have talked to people about getting tested. Why do they think they want to? And I have had a couple of other people to talk to, and I had a lot of fun. It was also really hard for me. Because you are standing around a lot of people who are dying...I mean, it's not the best to look at it this way but it is sort of the truth.

Daphne felt both comfort and sadness from the meeting, as she saw the people attending the meeting as “dying” because they had HD.

I saw them as dying because they had the disease. It's not the best outlook to have but it was sort of my perception of it that, you know, these people are going to die. I mean it was sad because some of these people are wonderful. Some of the people I met were just love, you know?

Daphne attempted to learn more about HD from attending the HD Society meeting but she experienced more than learning about the disease or getting support from people in similar situations. She projected the image of dying on the people at the meeting and found herself surrounded by her fear of her own mortality. As stated earlier, Daphne saw her own mortality in her father’s disease.

It is like having your mortality shoved in your face every day. It is like you’re going to die. You’re going to die. You’re probably going to die. Especially to a teenager it is really hard when you think you are, like, immortal, and then you have this shoved in your face every day: you are going to die.

When she attended the HD Society meeting this feeling became magnified and she left the meeting with a great deal of ambivalence. It was the first and last meeting that Daphne attended.

When Carla was ten years old a doctor talked to her about Huntington’s Disease and the fact that it was hereditary. This occurred at the time her mother was tested for the disease. She remembers being afraid that “my mom was going to die tomorrow.” By the time Carla was eleven, she was able to identify the symptoms of HD in her mother’s behaviour. She began to realize that she would not be able to live with her mother when her parents separated because of her mother’s symptoms.
I knew about Huntington’s. I didn’t know a lot about HD but I knew enough [about the symptoms] to know that I couldn’t really live with my mom. I knew that she couldn’t take care of me as well as my dad could. And I was thinking that in most cases the child goes to the mother. A lot of the times I was worried. I mean, I love my mom, but I knew she couldn’t take care of me.

Carla’s father told her friends and their families about her mother’s HD, and it upset Carla because she was not asked whether she wanted that information shared. She felt her rights had been violated, and she was angry that people had learned about her personal life without her consent. During the interview she explained that she does not talk about her mother or her mother’s symptoms with her friends. She will only talk about her mother’s symptoms if a friend is with her when she visits her mother, and then she only talks about it so that they will not be afraid of her actions. She finds that telling her visiting friends about the symptoms explains her mother’s inappropriate behaviour and protects her mother from judgment.

Emma’s mother told her that a disease “ran” in her father’s family, and that her father had taken a test to determine if he was “eligible” for the disease. Her mother told her that the disease “affects the nerves and stuff like that.” Emma’s mother had cancer and became seriously ill shortly after relaying the information about HD to Emma. Emma’s father “never really talked about it at all.” Unlike John, who used the Internet to gather more information about HD, or Daphne who went to a HD Society meeting, Emma uncovered more information about the disease by accident. After her mother died, she found “hospital papers” regarding her grandmother’s death from HD: “I found papers on, like, what it would do to you and stuff like that, and how to behave.”

When asked about what she told her friends, Emma replied that she made HD an “excuse” for her father’s behaviour.

I just say he’s got a disease, he can’t help the way he acts. He’s really embarrassing and he says the wrong things and he gives people the wrong impression. With the disease . . . I got to explain to people because then it makes me feel a little bit better that they know he’s not normally like this, maybe.
Emma, like Marilyn and Carla, was motivated to tell her friends about Huntington’s Disease because she was concerned that her friends would think that her father’s rude behaviour was normal. She did not believe that his behaviour was the result of HD, but acknowledged that his behaviour might be worse because of the disease. Having the label of HD helped relieve her of the social stigma of having an inappropriate father; it gave her an “excuse” for her father’s actions and helped her feel “a little bit better.”

Tessa, Margaret, and Kathy are sisters in a family whose mother has HD. Tessa was told about HD by her mother, and Margaret and Kathy were told about HD by both their parents. Each of the sisters had a different response to learning about the disease.

Kathy, the eldest sister interviewed, felt that she experienced the progression of the disease differently because she had known her mother when she was healthy.

Oh, definitely. The two younger ones – I’m sure they don’t have memories of her. So, they could go back twenty years and say, I grew up with a sick mom, where I can say, Well, she was healthy for a while and then she started going downhill. So, I kind of saw more of what happened to her as she deteriorated.

Kathy was ten when she was told about HD. “It just went in one ear and out the other.” She felt she was too young at the time to understand, and it was not until she experienced her mother’s symptoms that she began to understand the disease. When Kathy was thirteen she started having some emotional problems that she called “self pity.” Her parents sent her to counsellors but she wouldn’t engage in the process. “I wouldn’t talk to them. I was always telling my parents it was a waste of their money.”

Margaret had a different experience than her older sister. Margaret remembered her need to attend counselling immediately after receiving the information to help her understand “how to deal with it.” She struggled not only with her emotions but also with uncertainty about how to tell her friends.
I would have been ten. And I remember I went to counselling because I was upset. I didn’t know how to deal with it. I didn’t know how to tell my friends what she had, and I was really emotional and sensitive and wasn’t sure how to deal with it. So, I went to counselling for that. I remember I knew that my mom had Huntington’s Disease and I didn’t know what my chances were. I don’t know when I discovered that but I know for a fact that I hadn’t known that then.

Tessa, the youngest sister, was three years old when her mother became symptomatic. She has no memory of what she was told when she was ten, except that her mother said she was symptomatic at thirty-five. She did a large school project on Huntington’s Disease and came to learn a great deal about the disease in the process. She emphasized that she did the project herself and “didn’t ask the family anything.” The information about the disease allowed her to discriminate between her mother’s behaviour and the behaviour of other mothers. Additional experiential knowledge came with an increase in the severity of her mother’s symptoms.

All three of the sisters were hesitant to talk to their friends about their mother’s illness, and chose to confide in only a few close friends. All three sisters got additional information regarding HD from pamphlets that their father had in his study.

In summary, all of the adolescents could describe the disease factually, although Tessa, Margaret, and Kathy were not as descriptive as the other adolescents, and had a harder time relating the information they had. All of the adolescents were able to tell some of their friends, and they all included experiential knowledge of the symptoms that they witnessed in their parent as part of the explanation of HD. Marilyn and Tessa were motivated to tell their friends because their fathers’ behaviour was embarrassing to them (whether it stemmed from HD or not). They reported that there would be less of a stigma from HD than from their fathers’ actual behaviour.

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29 Tessa told me that she was told about her mother’s disease at three years old. As the interview continued, my questions helped her contextualize the story. This participation assisted in the clarity of the story and demonstrates the co-construction of the stories rather than the actual history.
As a therapist I am a professional secret-keeper. I am often the very first person to whom someone risks telling a long-held secret. Several decades of guiding those struggling with secrets have taught me that secrets have a remarkable, if paradoxical, power to unite people and to divide them. The power of secrets and the impact they have had on the adolescents in this study became evident as I questioned them about how and what they learned about HD. As stated in the beginning of this chapter, a number of adolescents had an intuitive knowledge that something was wrong in the family. Even if the children were not told of an illness or specifically about HD, they soon become aware of changes in the atmosphere at home and in their parent’s health and/or behaviour.

Family secrets can be destructive. Families are support systems. Our identity and ability to form close relationships with others can be attributed to the trust and communication we develop with loved ones. If family members keep secrets from each other – or from the outside world – there can be emotionally challenging consequences. In the narratives of the nine adolescents, secrets played a role in dividing family members, in discouraging individuals from sharing information with anyone outside the family, and in miscommunication within the family, causing unnecessary guilt and doubt, and creating special intimate relationships between a parent and an adolescent when a secret was shared.

Secrets that bind and divide

Secrets are not always destructive; they may serve to establish bonds between two people. When siblings keep secrets from their parents, for example, they attain a sense of independence and a feeling of closeness. But the creation of any secret between two people in a
family actually forms a triangle because a secret always excludes — and therefore involves — another (Imber-Black, 1998).

Four of the adolescents had a close connection — a bond — with the parent who shared information about the family secret. The information usually consisted of information about HD (who had the gene for HD, etc.) and that it was a hereditary disease. Three of the adolescents (Stephanie, Marilyn, Emma) had this special relationship with their mother, while one of the adolescents (Margaret) had a special relationship with her father.

Stephanie noticed something was wrong at age eight but she did not ask anyone what was happening to her father. “I had to figure out for myself. They [the family] keep everything hidden.” Stephanie was told about HD at age eleven when her mother presented her with a large binder full of information about HD. Stephanie had always felt her father was different from other fathers but she began to learn from her mother that the difference had to do with a disease in the family called HD. When Stephanie identified her aunt’s behaviour as HD her mother began to disclose other “family secrets” about her father and her father’s family.

Stephanie developed a strong alliance with her mother, and often used the plural “we” to include her mother when she spoke of her father and her father’s family.

We don’t visit my dad’s side if he’s not there. We really don’t like them . . . they’re really conceited and think they’re everything. They always want to know what we’re doing . . . how much money we’re spending.”

She also withholds information from her father until her mother decides to disclose the information.

My dad knows that I play hockey and that I do modeling. He didn’t know about my activities before, and my mom didn’t want to tell him. Cause he would have been, like, how much is that? [Said sarcastically] He’s really worried about money. I don’t know why. He stopped doing physio and stuff because there was a charge. He’s like, I don’t want to do that because it costs money. My mom told me about that.
A bond developed between Stephanie and her mother that had as its foundation the sharing of family secrets and communication about managing family members. Implicit in this relationship is the exclusion of her father and her father's family; thus, the triangle is formed with Stephanie and her mother bound by the secrets that are kept from her father.

Marilyn described her family as "secretive," and did not feel comfortable talking to anyone but mother. "She is pretty much the only one in the family that really isn't that way." During a Christmas dinner Marilyn "put everything together" concerning her aunt's behaviour. She asked her mother about her aunt and was told her aunt had HD. She did not approach her father or any other family member, as her mother is "the only one that tells me anything."

Marilyn did ask both her parents why they had withheld the fact of her father's condition from her.

Finally, I thought that I am just going to ask. My parents would always be kind of be whispering you know, but the truth is they weren't going to tell me. They were going to hide it as long as they could; possibly until I moved out when I was nineteen or so. I don't know how they would of.

It was her mother that answered saying, "we wanted you to get through your life without knowing." Marilyn could not believe that her parents thought she was "stupid" and wouldn't figure it out.

Emma's father did not tell her that he had been tested and that he had the gene for HD. Emma described his silence as an indication of a "secret sort of thing." Emma and her mother did not discuss her father's behaviour, or much about which family members had the disease. Like many of the other adolescents in this study, Emma had to figure things out for herself. Emma felt very close to her mother and trusted that she would communicate to her what she needed to know. Her mother, who was in remission from cancer, suddenly became ill and died from a brain tumour shortly after disclosing that her father had HD. She was unable to discuss HD with her father, and was unsure as to how to assist him in daily living. She found herself in a situation
where holding a secret about her father had left her unable to manage the relationship with her
father as she attempted to take over the role of caretaker of the family after her mother’s death.

Margaret was the only adolescent who reported a close relationship with her father.

My dad has always been the comforter in the family. My mom can watch
anything and she’s a very strong woman but she doesn’t cry at all. I’ve never seen
my mom cry, which is hard to believe considering she’s got HD, and if I had HD
I’d feel kind of a failure because I couldn’t do so many things. The reason I go to
my dad is probably because he makes things a lot more clear and he’s able to
express things more clearly than my mom could.

Three of the adolescents (Stephanie, Marilyn, Emma) developed or had a close
relationship with the parent who disclosed the information about HD. They perceived that there
was a secret, but didn’t understand why until they were told that HD was in the family and that it
was affecting a parent. At the same time, Stephanie, Marilyn, and Emma are not close to their
fathers with HD, and do not seek information about HD. They also do not disclose what is going
on in their own lives, or how they are feeling about HD or other general family concerns. It is
important to note that the three adolescents may never have felt close to their fathers. What is
apparent is that their fathers’ silence did not enhance their relationship and did not encourage
them to seek information from him.

Daphne and John were not told about Huntington’s Disease until they attended a meeting
with their family and a medical professional, when they were twelve and fourteen respectively.
They saw their father for the first time in a year at this meeting. Both Daphne and John learned
that their father had wanted to tell them about his disease but their mother was opposed to the
idea. Both adolescents felt that their mother had violated their right to know, and they were angry
that they were not informed and that it had been held as a secret. Daphne remembered being
angry.

I was really mad at my mother for not telling me because I would have rather
known right up front. I would have rather known the moment he found out. I
would have wanted to know right away because then I could have gone out and
done the research on my own. If I had any questions I could have found them out
myself instead of having to rely on everyone else to do it for me. You know? So, I mean I understood she had a good valid point but I would have rather had the opportunity to do it myself and to seek out help if I needed it or not.

John recounted,

For me, personally, I would rather have known right away instead of waiting around for six or eight months. I know there was mediation going on. I didn’t know about it at the time. I know about it now.

Both Daphne and John would have liked to have been informed of their father’s medical condition as soon as it was determined that their father had the gene for HD, which was a year before they were told. It was not until his father’s symptoms became apparent to John that he discussed HD with his mother. John does not talk about the disease with his father. Daphne has chosen not to talk about HD or being at risk for HD with her mother or her father. She does speak to her grandmother, who lives with her and her father.

Secrets that lead to misunderstanding

Children are keen observers of their environment, and studies of family disruption (such as divorce) have shown that they construct meaning for themselves about the disruptive event; even blaming themselves for problems and the changes that may ensue (Hetherington & Stanley-Hagan, 1999). As described earlier in this chapter, Carla believed that she might be responsible for her parents’ fighting and for the changes she saw in mother’s behaviour because she couldn’t understand what was happening. Withholding knowledge about HD from the family it affects can cause unnecessary guilt and doubt.

Given that adolescents can assume responsibility for problems in the family that they do not understand, they can, according to Korer and Fitzsimmons, be caused undue anxiety or doubt by misinformation about the disease (1987). The adolescents in this research did not perceive
misinformation as a problem; rather it was the perception that HD was a family “secret” that caused the adolescents to feel constrained, uncomfortable, or unable to ask questions about HD.

When a family’s secret involves a challenging situation like HD, day-to-day family relationships and interactions with the outside world are strained. The adolescents learned that one did not discuss HD in the family by observing family communication styles around talk about family members who had HD. Most of the adolescents did not seek information about HD as they felt it was “not ok” to discuss the disease within or outside of the family.

Confusion for the adolescents appeared most frequently around their misunderstanding about Juvenile Huntington’s, and their confusion around the issue of inheritability of HD. Carla best exemplifies this confusion when she talks about her understanding of her risk for HD after preparing a class speech on Huntington’s Disease. She looked at the literature on HD and determined from reading about Juvenile Huntington’s that she could become symptomatic at any time. It is important to note that she did not discuss her confusion with her father or mother.

I’m really, really scared. I mean especially the fact there’s a thirteen-year-old guy who has HD symptoms already. And I’m just, like, I thought this happened to forty- or fifty-year-old people. I was reading up on it, like, maybe last semester. I did it for a school project. I did a speech and I figured out it could happen to kids around two.

Tessa was also concerned about when she might have symptoms if she had the gene for HD. “I could get it soon or not soon. Isn’t there someone who got it who was like a teenager?” Like Carla, Tessa did not seek answers from her father, mother, or the social worker from the HD Society.

Marilyn is a good example of how confusion around the probability of carrying the gene can lead to erroneous conclusions.

They said basically that you have a 50:50 chance. ... I kind of heard that before but I always thought, ok, it’s either brother or me, I guess. ... But I don’t want my brother to get it, but I don’t want me to get it. Oh, no!
Because Marilyn felt she could talk to her mother, she asked her again about probability. We discussed probability further but then she sat me down and said, No, it’s 50:50 for you and Matthew. I’m, like, you mean we can both get it? You know. And they say, yes. It’s like, Oh no! But you mean we can both not get it, too? So, I never thought about it that way. I always thought about him or me.

These examples reveal the adolescents’ confusion with regard to Huntington’s Disease. Most of the adolescents had misunderstandings or unanswered questions about the disease. The keeping of family secrets around HD created an atmosphere in which most of the adolescents felt that the subject could not be brought up, even if only to ask for clarification of information about the disease. The consequence of this misinformation was that some of the adolescents had issues around inheritability and early onset that caused them anxiety and concern.

**Summary**

The preceding stories describe the experience of the adolescents’ process of learning about HD. Six of the adolescents began to intuit that something was wrong between the ages of five and thirteen. Their awareness that something different was happening was not related directly to HD and occurred prior to receiving information about HD. Something was wrong either with the behaviour of a parent or relative, the behaviour of the family, or because there was denial of access to a parent.

The experiences of these six adolescents reflect the process of insight that during childhood is essentially sensing. It is a pre-verbal intuition that signals to the child that something is wrong in their world. The six adolescents experienced this pre-verbal intuition as any child would. It was not an experience unique to HD in the family, but rather an experience that was the result of the impact of HD within their family, and the resultant behaviour of the adults in their family.
The experience of these six adolescents demonstrates that keeping a secret about HD from children does not necessarily mean that they do not know that something disruptive is happening in the family; nor does it alleviate distress. Stephanie, Marilyn, and Margaret knew that something was wrong but they did not ask for clarification from the adults in their family. They knew that it was somehow a family “secret,” and they carried the confusion with them rather than risk asking for information.

All nine of the adolescents were told about HD when they were between the ages of nine and fourteen. Various adults in their worlds told them about HD – an affected parent, a non-affected parent, both parents, an extended family member and medical health professionals in the presence of family members. There were two levels to learning about HD for all the adolescents. The first level was being provided with the factual information regarding the disease. The second level of learning came from the experiential knowledge of HD that resulted from living with a parent or relative with the disease. It was at this level that the adolescents began to see HD symptoms in a parent or relative. Both levels of learning about the disease allowed the adolescents to begin to discriminate what was and what was not a symptom of HD, and to discriminate between their parent’s behaviour and the behaviour of other parents.

All of the adolescents were able to describe HD in factual terms. When describing their affected parent’s symptoms to their friends they would use both factual and experiential information. Adolescents in general are concerned with how their peers judge them, and with what is considered appropriate and inappropriate behaviour to their peer group. The at-risk adolescents used descriptions of their parent’s HD-symptomatic behaviour as a way to explain behaviour that might be perceived as inappropriate by their friends. Marilyn and Tessa were motivated to talk about the symptoms of HD as a way to avoid the embarrassment caused by the behaviour of their symptomatic parent, whether or not these behaviours were actually a result of HD. In this case, HD provided them with an excuse for behaviour that may cast a judgment on
themselves or their families. Carla explained her mother’s symptoms only to those who spent time with her at her mother’s apartment. She was concerned that they would be afraid of her mother. She can be seen as protecting her mother by disclosure of her mother’s symptoms to her friends.

Secrets about HD and the hereditary nature of HD created uncertainty and confusion for the adolescents in this research. The adolescents who intuited that something was wrong in the family did not ask the adults in the family for clarification. As Carla said, “I had no idea what to ask.” Marilyn, Emma, and Stephanie recognized that the “secretive” nature of their families meant that they in turn were to keep secrets and not ask questions.

The sharing of the secret had the power to bind relationships between the adolescent and the teller of the secret. Four of the adolescents reported a close connection with the parent who disclosed the information about HD. Conversely, for three of those adolescents a more distant relationship was created with the parent who the secret was about – the parent with HD. They chose not to divulge details of their lives or their feelings about HD with the parent who had the disease.

Secrets also led to misunderstandings. The adolescents felt they could not ask questions about HD or talk about their concerns because they were not supposed to know about the disease. By watching the communication patterns of the family, the adolescents learned that it was not okay to talk about HD within the family, or to go outside the family to discuss HD. The consequence of this silence was that the adolescents carried incorrect information about inheritability and early onset that caused them anxiety and concern.

In naming the legacy of HD came a new set of challenges for the adolescents in this study. They were able to learn about HD, and came to identify some of the symptoms of HD in their parent or relative. The knowledge of the legacy led to a new set of complications and implications in the relationships of the adolescents. Learning how to negotiate relationships both
within the family and outside the family became more complicated because of the impact of both the factual and experiential knowledge. The potential impact of their symptomatic parent's behaviour on peer relationships added an additional stressor on the adolescents' lives.

In Chapter VI, an exploration of the impact of HD on the adolescents' relationships both inside and outside the family will be examined to further illuminate the experience of being an at-risk adolescent in a family with HD.
CHAPTER VI
EXPERIENCING THE LEGACY: HUNTINGTON’S DISEASE
IN RELATION TO RELATIONSHIPS

“I can’t remember my mom as healthy. I’ve never had a mom like everyone else.”

As a therapist, I have learned that most of my clients describe their wellbeing in relation to relationships. They are either happy or unhappy because they are in a relationship or because they are not in a relationship. Relationships play a central part in the wellbeing of adolescents. Social-emotional development during adolescence is characterized by changes in emotional attachment to parents and peers, and a gradual assumption of adult roles, responsibilities, and status. Typically, as the adolescent approaches adulthood, emotional dependence on parents decreases, and greater parity in the relationship occurs. This is accompanied by an increase in the time the adolescent spends with same- and opposite-sex peers, and the development of mature intimate relationships, usually with opposite-sex peers. Preparation for other adult roles and responsibilities, such as worker, parent, citizen, and spouse, also occurs during this period.

Purpose and Outline of Chapter

In Chapter V, the various ways in which the nine adolescents learned about and experienced Huntington's Disease were discussed. The influence of family secrets and the adolescents’ misunderstandings about HD were further explicated. The impact of this knowledge was reflected in the adolescents’ perception of and interaction with both family and friends. In this chapter, the effects of HD on adolescent relationships will be explored. The first section of the chapter will illuminate how the at-risk adolescents perceive and judge their parents. The following sections of the chapter examine the effect of HD on family relationships and non-family relationships, and the impact of HD on these relationships. Family relationships will
include the parent/child relationship, sibling relationships, and extended family relationships. Non-family relationships will focus on peers. The effect of HD on peer relationships is evidenced in two ways: fear of embarrassment around the affected parent’s behaviour, and fear of how peers will judge the parent or themselves because of the exhibited behaviour.

The Adolescent Perspective

Before examining family relationships, it is important to emphasize that adolescents do not necessarily use Huntington’s Disease as a paradigm to understand their parents’ behaviour. All the adolescents reported that their awareness of a difference in a parent was marked by changes in the parent’s movements or behaviour. This awareness was gradual, and part of an unfolding process that occurred over time. The present research caught a slice in time of the adolescents’ understanding of HD, and this understanding will continue to evolve over time. For the adolescents in this study, the knowledge that a parent had the gene for Huntington’s Disease added a new dimension to their view of their parent but did not become the primary “lens” through which the parent was viewed. Problems were not always related to HD; for some of the participants problems were based on issues that predated them learning about HD in the family.

Emma remembered always having difficulty with her father’s anger and how he behaved around people: “He’s really embarrassing and he says the wrong things and he gives people the wrong impression. He has been like that forever.”

Daphne spoke of her struggle with her father around a sense of abandonment after he left the home when she was three. She could not understand how her father could leave and come back years later and expect to be her father.30 “Why should I take him back when he left?” She

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30 Daphne spoke of her father leaving when she was three and returning when she was “eight or nine.” When doing the lifeline she spoke of not seeing her father for one year when she was thirteen years old, and of seeing him again when she was fourteen at the meeting where she and her brother were told that their father had the gene for HD.
still tries “really hard to forgive him.” These issues with her father predated knowledge of his illness and continued to be a primary focus for her even after his disease status was revealed.

It may be that through the adolescent’s eyes it is the identity of “parent” and the expectations associated with that role that creates the lens through which the parent is viewed, whether or not they are the parent with HD or they have symptoms associated with HD. Marilyn demonstrates this when she laments the fact that her father is not fulfilling his parental role.

And my mom said the saddest thing for her is that he has the time but yet he just decides to sit and watch TV rather than spend time with us. And I think that in a way that’s true. That’s what makes me so mad is that he just doesn’t seem to put the effort in to be the father that we need.

The nature of the parent-adolescent relationship does not depend only on what happened in the relationship during adolescence or on the experience of HD symptoms. The co-construction of the stories between the adolescents and myself gave the adolescents time to reflect on their memories, and helped provide an understanding of the events.\(^\text{31}\) For example, Marilyn felt that she never had a close relationship with her father, regardless of HD.

And now, like he’s at home pretty much every single minute I’m there. It’s pretty hard because I’ve never really liked him. It’s not that I don’t like him but I’ve just never had a good relationship with my dad. So, now he’s changed so much that it is almost like living with another man.

For Marilyn, her negative view of her father preceded the onset of his symptoms – the symptoms only exacerbating an already challenged relationship.

Given that the average age of onset for Huntington’s Disease is between thirty-six and forty-five years\(^\text{32}\) there is a likelihood that parents will become symptomatic around the time their children become adolescents (Hayden, 1981). It is possible that problems in the parental-

\(^{31}\) The co-construction of the history of HD in the lives of these adolescents created an understanding of the events rather than an actual history of the events, i.e., the age and cognitive development of the adolescent influenced their understanding of the events.

\(^{32}\) Although the typical age of onset is between thirty-six and forty-five, the disease may remain quiescent until the individual is well into old age.
adolescent relationship come to be viewed by the parents as HD-related when in fact they are based on issues surrounding the typical adolescent-parent relationship.

John and Daphne knew nothing of HD, and had not known anyone who had the disease before the family meeting with the mental health professional. The meeting occurred when John was twelve and Daphne was fourteen. Even though they learned about the disease and its symptomatology at the meeting, they continued to judge their father based on previous issues around abandonment, lack of anger control, his unemployment, and his inability to rise above his depression. Again, despite the information about HD and the witnessing of choreic movements, John and Daphne continued to view their father in the role of parent rather than using HD as an optional explanatory tool. John spoke about his father’s unemployment as a part of his father that he disrespected.

In the beginning I think that I visited him more. And then I kind of leaned off a bit because I didn’t want to hang out with him as much anymore. ... I’ve known my dad forever and he has never had a job. I started losing respect for him for that reason. The longest job I think he has had, I think, was sort of like four months. It was a gas station job.

He judged this aspect of his father as a flaw in his character even though he acknowledged that his depression was associated with his illness. He remained unsympathetic to his father’s symptoms because of his father’s past behaviour.

I think I was seventeen and he stopped going anywhere and he got really depressed. He probably had a reason, but he stopped going outside anymore. He use to go on drives, I think. He would go 4-wheeling and stuff, and he stopped doing that after a while. And then he would be inside on the computer all day. It was really pathetic.

Daphne spoke of her father’s depression much as her brother did. She, too, lacked empathy for his behaviour, even though she acknowledged that it might be caused by Huntington’s Disease.
You know I never could understand how he could sit around the house all day when he knew that the end was coming. I understand that he has a depression. So do I. Right? But still, I would have gotten out in the world and done something not sat around in the house all day.

She also judged her father’s anger based on past behaviour and not as a symptom of his disease.

I’ve always been a little bit scared of him. I can trace it back to one memory. [Laughs] Yeah, it is really funny that I can do this. It’s a long time ago – but it stuck with me, and I have always been scared of him. He was really mad because someone didn’t show up with the keys and he fucking ripped his shirt off. Ripped his shirt and the buttons off and stormed around and yelled and screamed and stuff. I was little, like, I was little bitty and you know, it scared me. And I have always been a little afraid of him. And because I have never really spent enough time with him I don’t know how he is going to react.

It may well be that due to the limited time she spent with her father when she was growing up she was unable to understand or predict his behaviour. She had experienced a traumatic event around her father’s anger when she was a little girl. The event created a fear of her father’s reactions, and, consequently, informed her explanation of his behavior; she did not use Huntington’s Disease to explain his anger.

As reflected by the co-participants’ experience of their symptomatic parent, the nature of parent-adolescent relationships does not depend only on what happens in the relationship during adolescence. Relationships with parents over the long course of childhood are carried forward to influence, at least to some degree, the nature of parent-adolescent relationships. The adolescents in this research judged their parents based on a lifetime of experiences with their parents. Knowing that a parent was symptomatic and that some behaviour may be associated with HD was not always pertinent to how the parent was viewed.
Family Relationships

Parent/Child Relationship

Parents are prominent figures that have a great influence on the adolescent. The significance of adolescent-parent relationships seems to be mediated by the adolescent’s gender and age, and by the parent’s gender. Adolescents tend to view mothers as the “emotional core” of the family – supportive and receptive – and fathers as the “crisis mangers,” providing more instrumental help (Frey & Rothlisberger, 1996). Most of the at-risk adolescents in this study viewed their parents in similar ways. Carla reflects this view of her symptomatic mother when describing how she comforted Carla.

When she was in the hospital I remember the first time I was there. I was crying. She’s lying in a bed. I came down and lay beside her. I just started crying. She gave me a hug and I started crying. She’s like, don’t cry. I’m fine. I’m going to be better.

Viewing mother as the emotional core may be the consequence of the type of conflict that the adolescents were privy to, which typically revolved around issues of daily living, and often involves the adolescent’s mother. Four of the nine adolescents (Carla, Tessa, Margaret, Kathy) have mothers who are symptomatic. Three of the four adolescents have been with their mother when they were not symptomatic, and recalled times when their mother took care of managing family life. As cited in Chapter IV, Margaret was the only adolescent of the nine adolescents in the study who viewed her father as the “comforter” and the one she went to for help with her problems.

It may well be that in families where a parent is symptomatic, the responsibilities for daily living fall into the hands of the non-symptomatic parent, whether it is the mother or the father. Margaret perceives that her father is the parent that is capable of the emotional support she needs. Stephanie describes her mother as playing both parenting roles.
It’s usually me and my mom who end up doing more. She takes up the time when he doesn’t spend time. Like, she takes us both. She takes me to the hockey games, which probably a Dad would do, and she takes me shopping. She does them both. Like, my mom’s the Mom and the Dad.

Marilyn struggled with her own angry outbursts directed at her father. In her attempt to understand her behaviour she turned to her mother for help. She perceived that her mother could help her understand and begin to identify possible reasons for her frustration with her father. In this case, Marilyn’s mother was not only an emotional support but also a provider of social verification about her own disappointment with her father’s actions. When discussing her frustrations with her mother she was able to make sense of her own anger as well as realize that her mother was “sad,” and that she was not alone in experiencing difficulties with her father.

The at-risk adolescents generally perceived their fathers, whether they were affected or non-affected, as more judgemental and less willing to negotiate with them. Margaret’s sisters found their father (the non-affected parent) to be more distant and judgmental than Margaret did. Tessa felt quite removed from her father, and found him unavailable: “Dad is home on the weekends, pretty much. But he’s outside fixing the car, or in the basement, or working on his computer or something. No one goes to my field hockey games.” Kathy found him less willing to negotiate with her: “He was the only one who didn’t want me to go because it cost money to go on vacation. Everyone else, Mom too, she’s like, Go along, have a ball. . . . Not Dad.”

The at-risk adolescents in the same home had very different experiences of the family environment, and, in particular, of how they were treated by their parents. Birth order of siblings played a role in how the adolescents experienced their parent with HD. Kathy knew her mother before she became symptomatic, while her sister Tessa has no memory of her mother without the symptoms of HD. Growing up with a mother who was non-symptomatic created a different experience for Kathy, as she acknowledges when talking about the progression of her mom’s disease.
It’s kind of sad to watch someone deteriorate like that, especially over a long period of time. . . . Mom’s had it for twelve years. That’s got to be harsh. . . . and my little sisters wouldn’t have any memory of my mom when she was healthy.

Changes in parenting roles can also affect the relationship with the parent who is symptomatic. Marilyn’s father had been busy creating a career, and was seldom home or involved in her everyday life. When he became symptomatic and remained at home, it became difficult for her to have a good relationship with him as she did not have one prior to his disease.

John was the only male adolescent in the study. His father was symptomatic, and John did not speak to his father about HD. Adolescent males tend to talk more with their fathers than with their mothers about general interests and problems, especially sexual problems. John’s parents were separated at the time he learned about his father’s illness. John’s lack of communication with his father may have been a result of living separately from his father rather than owing to the impact of his father’s illness. Although he did not speak to his father about his concerns or about his at-risk status, he was able to speak to his mother a year after he learned of his father’s disease.

Yeah, I did talk to my mom but it took me a little while before I did. My mom wanted to talk to me about it but I kind of wasn’t ready. . . . I don’t think it actually hit me that it could really affect me. I was just kind of worried about my dad and that he was going to die pretty soon. I still had the image that it was going to be a lot quicker that what they told me. So, I started talking to my mom I think around thirteen. I started talking to her about what was actually going on in my head and stuff like that. And I started noticing – I actually started noticing – twitching. [Spoken softly]

The adolescents in this research were exposed to the intellectual deterioration, choreic movements, and personality changes of their HD-affected parent. All the adolescents reported that their awareness of a change in their parent was marked by changes in movement or behaviour. Yet, as stated earlier, Huntington’s Disease was viewed as only one aspect of their parent.
For the adolescents in this study, HD was perceived as a problem, but not the only problem. While domestic violence, alcoholism, and sexual aggressiveness have been linked to HD, such problems also affect many families without a genetic disorder in the family. Some of the adolescents believed that the problems they experienced predated the onset of the disease. Emma reflects this understanding even though her social worker sees the problem as related to HD.

There's still a lot of yelling, and he's still got that temper. The social worker tells me that it is HD, but I don't believe it because she keeps telling me and assuring me that it's from HD, but I don't think it really is... Some of it might be, but you can't use that as an excuse all of the time.

To reiterate, it may be that the adolescents view the identity of "parent" as more crucial and relevant than that of "parent with Huntington's Disease." The expectations of parenting create the context in which the parent is judged, and the disease becomes secondary.

It can be difficult for the adolescents to discriminate between behaviour that is a reflection of Huntington's Disease and behaviour that is not a part of Huntington's Disease. Marilyn not only reflects this process of discrimination but also demonstrates how she tries to determine what is HD and what is her parent. She talked about how her father does not help her mother around the house because he feels he "can't do it." She believes that it contributes to her dislike of her father because "he uses the disease to his advantage."

Marilyn fights a great deal with her father, and her mother tells her that she is frustrated by her angry behaviour. Marilyn acknowledges that she does argue with her father too much, but finds his reactions to their arguments necessary; they demonstrate to her that he is "not normal." She believes that only when she "pushes" him into anger does she accept that he has HD and that he really can't help how he behaves.

Regardless of the perception that the adolescents had regarding their mother or father, they tended to judge their parents by the same standards that any other adolescent would use
(Collins & Collins, 1990). Huntington’s Disease was not always relevant to how the parent was viewed, and many times it was not a part of the criteria they used to judge or understand their parent.

Sibling Relationships

Of the nine adolescents in this study there were two sibling groups. John and Daphne are brother and sister whose father has HD; Tessa, Margaret, and Kathy are sisters whose mother has HD.

When I asked John if he spoke to his sister about their dad’s HD he replied, “After that meeting, we talked about that, and we talked about Dad’s letter and stuff like that.” When I asked John if he and Daphne talked about their risk status he replied, “We didn’t say much. I don’t think either of us ever talked about that.” John seldom mentioned his sister in his interview, and described her as an “emotional” person. The person he confided in was his mother, and he spoke of having a different relationship with his father than his sister did.

Daphne, on the other hand, spoke about her relationship with John in different terms. Johnny and I are very close. Like, we’re really, really close. So, that’s cool. I mean the one thing that we have always had is each other through all this craziness. We’ve handled stuff together and that is the basis. We have always been like that and we have always been really close at school.

Daphne, who is the elder of the siblings, perceived a close relationship with John. John did not speak of his relationship with his sister as “close.” I also found the same situation when I was interviewing Tessa, Margaret, and Kathy.

Kathy, the eldest of the three sisters in the study, feels that she has “parented” her sisters since she was fifteen.

Mom and Dad trusted me to baby-sit them a lot and, you know, they tell me quite a bit. We talk about the [father’s girlfriend] situation and all that kind of stuff. So, we’re pretty open with each other.
Tessa described her relationship with her sisters as "not close." "We just don’t talk about it. So, we don’t talk about anything really." Margaret’s response to how the family feels about her mother’s HD is similar.

I don’t talk much. I don’t talk with them. See, the thing is people say, oh, you should talk with your sisters, try talking to them, talk about Huntington’s Disease, and that’s the only thing I hate talking. I’d just rather not deal with it because I have my own way of dealing with it and I don’t like sharing my feelings with other people.

In both sets of siblings the eldest child felt that there was closeness in the sibling relationship while the younger siblings did not feel close. The people that Tessa and Margaret felt the most comfortable talking with were their friends. Each of them had one or two special friends in their circle that they turned to when they felt the need to talk about their feelings. John found comfort in talking to his mother about his relationship with his father and his concerns surrounding HD.

Marilyn and Emma each have younger brothers. Marilyn felt that she and her brother did converse about HD in the family, even though he would deny talking about it “We did [talk about HD], and he says we didn’t, but we did a lot. And we always kind of knew Dad had it.” She also wanted to protect her brother from finding out about HD from their parents on his birthday.

I didn’t want them to tell him on his birthday because we had a big party and all that, because I found out just before mine. So, they told him about it a few days after it. And he’s so fine with it. I don’t know if he just didn’t understand or anything because he’s just like, ok.

As the interview continued, Marilyn talked about her brother realizing that maybe it was not “ok.”

Later on it got worse, like kind of, this sucks! But at the time I looked at him and thought, come on. Where’s your reaction? You know, he’s just like, fine. I don’t really have one.
Emma does not have a close relationship with her brother, and feels that she has had to carry the responsibility of her father’s illness herself.

No. No. He doesn’t do anything . . . He is so involved with his friends it is unbelievable. He doesn’t care about this stuff. He doesn’t even want to know about it. He doesn’t care. And he doesn’t care about nobody but his friends. Like, I mean you won’t get him to come to any of these meetings. He hates talking to the social worker. He doesn’t hate her. He hates talking to her because they talk about stuff that he didn’t want to talk about.

Emma does feel that her brother cares about her, but only because she learned that he missed her when she was away for a month. At the time of the interview, Emma was planning to move out of her home with the help of her social worker, and she felt that her brother might miss her.

Stephanie and Carla do not have siblings. Carla stated in the interview that she wished she did have a sibling so that she would not be so “alone” in dealing with her mother’s HD.

Extended Family Relationships

Stephanie, Carla, and Marilyn had relationships with grandfathers who had HD, with Carla having the closest relationship. All of them remember their grandfather as symptomatic. Carla and Marilyn visited their grandfathers in the hospital and learned that their grandfathers had a disease that was called HD. None of the three adolescents were told that the disease their grandfather had was hereditary.

John, Daphne, Tessa, Margaret, and Kathy had never met a family member with HD and knew nothing about it until their parent became symptomatic.

The adolescents seldom spoke of their extended family members. Their relationships were usually described around how the extended family members joined in the silence about their shared genetic heritage. As Marilyn states,

They knew before me. My whole family knew. The only person that
would not accept it was my grandma, she’s like, no, you don’t have it. He [her father] would say, yes I do. She was in such denial for months and months. Finally, she accepted it.

Marilyn, Stephanie, and Emma had aunts who showed signs of being symptomatic. Each of them guessed that their aunt had HD. As Marilyn recalls,

The whole family started talking about it a lot. I started listening a lot more. My parents gossiped – well, it’s not even gossip, it’s just like, should we tell her? I think she has it. You know, blah, blah, blah. They didn’t really think, like, that Marilyn was there.

Stephanie:

I can just tell. Like, it’s just because she was over at our house once and . . . she’s, like, kind of moving and her hands are going and feet kind of. Her mouth would be open and she would talk different . . . . My mom told me. Well, you could kind of tell it was like Huntington’s.

Emma:

Well, my dad’s youngest sister has it but she doesn’t talk about it. She tells people she doesn’t have it but we all know she does. What I see in my dad I see in her.

John’s and Daphne’s grandmother moved from the East to help take care of their father, who was beginning to have difficulty living on his own due to the symptoms of HD. John does not have a close relationship with his grandmother, and does not spend time with her. Daphne, on the other hand, lives with her father and grandmother, whom she feels close to.

Nana’s the best. And you know having somebody to make sure he’s ok and take care of him is actually what he needed. I mean he may not feel that way but anyone who knows, knows that that is what he needed. He needed his mom. You know?

Of all the adolescents in this research, Daphne’s grandmother was the only extended family member to provide information about the history of HD in the family. The information was of a very private nature concerning Daphne’s parents.
I know that Nana was told to not have kids. I know that Mom and Dad were told not to have kids. Well, Nana told me about her, and Nana also told me about Dad ‘cause she had warned him and said, look, this runs in your family. All these people are dying and yada, yada, yada. I don’t think you should have kids or at least go get counselling before you do. I mean . . . they actually thought about aborting me. You know, they have never regretted not aborting me. They love me but, you know, there was a point when they thought about it. They were so young then they actually couldn’t. I think they asked for it but she was too far along.

Daphne’s grandmother also plays a role in how Daphne sees her future.

Can I really be a Nana? Because she was warned not to have kids and she has to watch her son die – slowly – and die of Huntington’s Disease. Do I really want to put myself in that kind of a position? And is it really fair to the child to be put in that kind of position?

**Non-Family Relationships: Peers**

For adolescents, friends and friendships are central to their development of self-esteem. The adolescents in this research reflected many of the concerns that young people share when developing peer relationships. The narratives revealed that having a parent with HD affected peer relations in essentially two ways: through embarrassment caused by the affected parent’s behaviour, and through concern about how friends would judge the parent or themselves.

Having a parent with HD affected how the adolescents behaved within their friendships; it did not seem to affect who they chose as friends. All of the adolescents chose friendships as any other adolescent would, based on proximity and similarity in interests, for example. The differences emerged when the parent with HD became symptomatic, and the adolescents became uncertain about how the parent would behave. This uncertainty resulted in a perceived vulnerability; the adolescents were concerned for their parent with HD and/or for themselves. The adolescent was embarrassed by the parent’s symptoms, and/or feared the possibility of judgement by peers of either their parent or themselves.
Embarrassment

I just say he’s got a disease, he can’t help the way he acts. Because he’s really embarrassing, and he says the wrong things and he gives people the wrong impression.

Five of the adolescents in this study did not want to bring friends to their house as they were uncertain how their parent with HD would behave. Emma, John, and Marilyn were embarrassed by their parent’s behaviour, and felt that their friends would judge them poorly as a result of their parent’s behaviour. Emma was quite clear about not wanting friends at her house: “I try to keep my friends away from my home.” As Marilyn states, “I try to avoid any contact with other people and him. That’s why I never bring friends over anymore. It’s just too awkward.”

Margaret and Tessa, who are sisters, did not bring friends home because they were concerned about how their mother would be treated. Margaret remembered an incident when her family had just moved to a new town and her mother fell down the stairs in front of two new friends and their mothers. When she arrived at school the next day, her schoolmates were talking about her mother being “drunk.” She knew that her mother was not drunk. The incident sensitized Margaret to the possible responses her mother might receive from her new friends and their families. She became protective of her mother and only infrequently brought friends to her home.

Tessa did not bring friends home because she was concerned that her mother would be ridiculed, and she wanted to protect her.

I don’t bring new people over usually. I don’t know... I don’t like really talking about it. And some people just look at her funny and stuff. Well, like some people are really mean and they make fun of a lot of people and stuff, so I’m scared that they are going to.

Kathy, who is older than Margaret and Tessa, felt differently about friends visiting.
Most of my friends were ok with it. I had a few close friends and they enjoyed talking to my mom just as much as I did. I wouldn’t be embarrassed inviting somebody over to the house to sleep over. But there are some people, like, when you’re walking down the street, they’ll kind of glance at her twice, you know.

As stated in the last chapter, Marilyn and Emma were both embarrassed by their father’s behaviour. Emma felt her father behaved inappropriately in his comments and his anger. Marilyn felt that her father acted effeminately and would be perceived as “gay.” Both adolescents felt more comfortable blaming their father’s behaviour on HD rather than seeking other explanations for the behaviour.

Daphne described bringing friends home in divergent ways. She initially spoke about bringing friends home and being able to feel that her father was “cool,” and that her friends also liked her father.

Lots of my friends totally like my dad. You know, my dad is pretty cool. They totally like him, and lots of my friends spent a lot of time over at that house and he was great about it.

She did not feel embarrassed by her father’s symptoms. For Daphne, a criterion of friendship was acceptance of her father. “It’s not about Huntington’s Disease, you know. They have to accept him the way he is. If they don’t then they can go jump somewhere.” Daphne also spoke about bringing a friend home with her “for protection.” She realized that she had “always been a little afraid of him.” This fear was the result of uncertainty. “I have never really spent enough time with him, I don’t know how he is going to react.” Daphne described an incident in which she went to see her father to collect child support for her mother.

The first time we went over because they weren’t paying child support anymore. Mom didn’t want us to see him – we didn’t want to see him – until he started paying child support. So, I was the one that had to go over again. [Laughs] My fate! And I took my buddy Kenton with me. And he is like my best friend. We went over there together because the times before I have taken my friend Katie. Because I’m never quite sure how he is going to react. You know? They know why they are there . . . if he hit me, or something, then they can do something about it. Because I was never sure how he was going to react.
It is interesting to note that Carla also felt that bringing her friends home would protect her. In Carla’s case, she was looking for protection from her mother’s verbal outbursts and the potential conflicts that would arise between her mother and father.

My friends and their parents were afraid. Some of them are afraid to come to my house. But I’ve had a couple friends that are like, yeah. They’re like, oh no, she’s not dangerous. We know what can happen. . . . She’d never do that while my friends were around.

John felt uncomfortable about bringing friends with him when he stayed with or visited his father. “I did not bring friends as much. I would have one friend over with me.” He did not tell his friends about HD. “I never said anything for a long time about my dad.” What precipitated his disclosure to his friends was an event that occurred when he was in town and walking down the street with friends.

When all my friends first met him they didn’t actually meet him. We were at a birthday party together and we were going to a movie and we saw him walking down the street. And I gave him a big hug and my friends went, Is that your dad? They thought it was some drunken guy that was walking. Yeah. My dad’s not drunk. He has Huntington’s.

Five of the adolescents in this study were embarrassed to bring friends into their homes. Marilyn, John, and Emma were embarrassed because they felt their affected parent would behave in ways that would cause their friends to judge them differently. Two sisters, Margaret and Tessa, did not bring friends home because they were concerned that their mother could be hurt by their friends’ reactions. Kathy, the eldest sister interviewed, did not feel the same and felt comfortable bringing friends home.
The adolescents in this study reflected their concern for how they would be judged by their peers in the decisions they made around who they chose to tell about HD. Most of the adolescents began to disclose to their selected friends during adolescence, after they began to notice symptoms of the disease in their affected parent. For most of the adolescents the disclosure occurred between the ages of thirteen and fifteen. Stephanie recalls that she started “really talking” at around thirteen years of age. John was able to remember when he began to disclose based on when his father moved from the East Coast to be closer to John and his sister: “Probably late grade nine . . . Yeah, it must have been grade nine because we were only apart for a year.”

Four of the adolescents (Stephanie, Carla, Tessa, Kathy) confided only in their best friends. Stephanie and Carla had a number of close friends that they felt comfortable talking with. Both Kathy and Tessa would only speak to their best friend about HD. As Tessa explains, “Actually, I never talk about it. There’s really only one I could trust not to tell anybody. . . . She has been my best friend since grade four.” Kathy recounted, “Oh, I’d just say mom was sick. I wouldn’t go into an explanation. [With] . . . my best friend I would. But I wouldn’t sit down and tell everyone.”

Marilyn and Margaret confided in their boyfriends. As Margaret explains,

I’m very private. I don’t like to tell too many people about my situation. I only tell . . . my best friend and my boyfriend. They are the only people who know, really, what is going on. I tell my best friend a lot of things, but I don’t tell her as much because my boyfriend understands more and she’s kind of more judgemental. Craig can always tell when I’m upset, and it’s just really nice to be able to phone somebody and him being able to tell I’m upset and he won’t stop asking me until I tell him. Which sometimes isn’t good because I don’t want to talk about it. And if

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33 Research indicates that the need for intimacy intensifies in early adolescence and motivates teenagers to seek out close friends (Sullivan, 1953).
I say I don’t want to talk about it he’ll just back off. But, usually, I tell him because I want him to know. He doesn’t tell anyone my secrets.

Emma was the only adolescent who discussed wanting a friend who was not also a confidant.

Like, Lisa. I want Lisa just to be Lisa. I don’t want her to be my little counsellor girl or anything like that. I just want one friend who doesn’t know a lot, and somewhere I can just go and have fun and don’t have to worry about anything.

Although most of the adolescents began talking about HD in their early teens, when their parents became symptomatic, each had their own comfort level with regard to disclosing their experience of HD in the family. All of the adolescents voiced some concern about being judged by their peers because of their parent’s behaviour, and were therefore selective about who they chose to disclose information to. Four of the adolescents were comfortable speaking to their best friends. Two of the adolescents felt they could only confide their personal concerns to their boyfriends. Margaret also confided in her best friend, but felt she got more comfort from confiding in her boyfriend. One adolescent selected a friend that she would purposefully tell nothing to so that she could “just go and have fun” without concern about the judgment of others.

The adolescents in this research reflect many of the concerns that young people share when developing peer relationships. The impact of having a parent with HD on peer relationships emerged in the narratives in two ways: embarrassment of the affected parent’s behaviour, and concern about how they or their parent would be judged. It appeared that many of the adolescents were able to select friendships that they perceived as supportive, and that allowed them to engage in regular social functioning within their peer group. Having a parent with HD was an additional stress that many of the adolescents learned to cope with – some more effectively than others.
Summary

The at-risk adolescents in this study perceived and judged their parents as adolescents generally perceive and judge their parents. Mothers were viewed as the emotional core of the family whether symptomatic or non-symptomatic. Margaret was the only adolescent who viewed her father as the “comforter.” The adolescents perceived their fathers as more judgmental and less willing to negotiate, which is similar to adolescents in families where there is no parental illness. Birth order and/or changing parental roles in the family could have influenced the adolescents’ relationship with their parents. Although the symptoms of HD can affect a parent’s capacity to function in a parental role, the adolescents responded to their parent with HD in ways similar to how they had responded prior to a display of symptoms or a recognition of symptoms of HD.

As reflected by the co-participants’ experience of their symptomatic parent, the nature of parent-adolescent relationships does not depend only on what happens in the relationship during adolescence. The adolescents’ relationship with their parents has evolved over the long course of childhood. The history of the relationship carries forward to influence, at least to some degree, the nature of the current relationship. Knowing that a parent was symptomatic and that some behaviour may be associated with HD was not always the way the parent was viewed, and many times it was not a part of the criteria used to judge or understand the parent.

Of the nine adolescents in this study, there were two sibling groups. In both sets of siblings, the eldest child described relationships with siblings as close. The younger siblings described relationships that were not close and where they did not communicate with their siblings. Marilyn and Emma had younger brothers who were not a part of the research because of their age. Marilyn discussed HD with her brother while Emma had little communication with her
brother and did not talk about HD with him. Of the two adolescents who had no siblings, Carla voiced that she felt “alone” and wished she had a sibling she could talk with.

There was little discussion of extended family relationships in the interviews. Stephanie, Carla, and Marilyn had a relationship with their grandfather who had HD. Marilyn, Stephanie, and Emma spoke of relationships with their aunts, although it was only in terms of their aunts’ disease symptoms. Marilyn had several aunts, but the only aunt spoken of was the one who gave away the secret of her father’s illness. Since information about HD and who may be symptomatic for HD was kept secret, it may well have influenced the adolescents’ relationships with extended family members. Knowing that the family had secrets that were not shared with some family members or to people outside the family may have signaled that communication was generally not open to the younger members of the family. The implicit structure of communication in the family may have hampered the development of relationships with extended family members.

Marilyn and Daphne were the only adolescents who spoke of their grandmothers. Marilyn talked about her grandmother’s anger and denial of her son’s [Marilyn’s dad] illness. Marilyn experienced her grandmother’s behaviour as a block to communication. Daphne, on the other hand, lived with her grandmother and learned a great deal about her family’s history through her. She appreciated the role her grandmother played in her father’s life, and it was her grandmother’s presence that allowed her to live with her father again. In many ways, Daphne’s grandmother played the role of “comforter” and “social verifier.” Daphne was not able to find this relationship with her own mother. Her grandmother also confronted Daphne with the possibility of a future as a caretaker of a child with HD. It was a perspective that created a great deal of concern for Daphne.

The effect of HD on peer relationships was exhibited in essentially two ways: fear of embarrassment around the affected parent’s behaviour, and fear of how peers would judge the parent or themselves because of the exhibited behaviour. Most of the adolescents were able to
select friendships that they perceived as supportive, and that allowed them to engage in regular social functioning within their peer group.

The experience of the adolescents living in families with HD presented a picture of adolescence that was not dissimilar to adolescents who do not have a parent with a chronic debilitating illness like HD. The adolescents revealed that HD created an additional stress in the parental relationship that many of the adolescents in the research learned to cope with. Some of the adolescents were able to negotiate this relationship more effectively than others.

The adolescents’ experience of living in a family with HD was reflected in the way they learned about HD, the process of how they understood HD both factually and experientially, and their relationships with family, extended family, and friends. The impact of realizing that they are at 50% risk for HD is an area that none of the adolescents had spoken about before the interview. The adolescents were engaged in a process of understanding the impact of being at risk for HD that evolved as their own self-understanding evolved. The impact of this knowledge and the intersection of this knowledge with the tasks of developing a future direction after secondary school could have a profound effect on their own identity and choices for their future.

In Chapter VII the impact of HD on self-understanding and future self will be explored in order to illuminate how being at risk for HD impacts adolescent identity development.
CHAPTER VII
INTEGRATING THE LEGACY: AT THE CROSSROADS OF SELF AND FUTURE

SELF

“Who are you?” said the Caterpillar. Alice replied, rather shyly, “I - I hardly know, Sir, just at present - at least I know who I was when I got up this morning, but I must have changed several times since then.”

Alice in Wonderland, Lewis Carrol, 1865

Adolescence has been characterized as a time when individuals begin to explore and examine psychological characteristics of the self in order to discover who they really are and how they fit in the social world in which they live. What is important about identity development in adolescence, especially late adolescence, is that for the first time physical development, cognitive development, and social development advance to the point at which the individual can sort through and synthesize childhood identities and identifications to construct a viable path toward adult maturity.

Purpose and Outline of Chapter

In the first section of this chapter, the dimensions of self-understanding will be examined to explicate the complex process of identity formation and to explore the impact that being in a family with HD has on identity development. As Richards stressed, “There is an urgent need to know more about how growing up in a family with a history of genetic disease shapes an individual’s self-identity and relationships with others (both inside and outside of the family)” (1993:10). It will be demonstrated that the adolescent’s experience of being at risk for HD is best characterized as an individual process that is influenced by cognitive, developmental, and the socio-cultural context of the adolescent’s life. The narratives of the at-risk adolescents will be
used to illustrate these dimensions, and to help establish how the experiences in their family and social lives contribute to their self-understanding and understanding of future self.

The second section of the chapter will examine how the adolescents view their future, personal meaning of at-risk status, and views on pre-symptomatic testing. The view of the future will be explored using four themes that emerged from the adolescents’ experience: quality not quantity; staying in the present; future generations; it’s not the HD. The adolescents’ perspective on their at-risk status and testing will be discussed using four themes that emerged from talking about predictive testing: desire for future knowledge; fear of future knowledge; uncertainty about future knowledge; indifference about future knowledge.

**Dimensions of Self-Understanding**

Self-understanding is the adolescent’s cognitive representation of the self – the substance and content of the adolescent’s self-conceptions. The development of self-understanding in adolescence is complex and involves a number of aspects of the self. Examining adolescent self-understanding as it differs from child self-understanding can help elucidate this complexity. Children from the ages of seven to eleven or twelve are egocentric thinkers who are convinced that their beliefs are correct. They do not understand that beliefs are the result of a thinking process and can be arrived at differently. They assume not only that their beliefs are correct, but that they are the only beliefs that can be held and that all people share them. As children grow toward the teenage years, they begin to think in a different egocentric way. At about age eleven or twelve, children begin to realize that beliefs are the result of a thinking process. They also come to understand that other people think, too, and that their own beliefs on a certain subject may differ from those of others. Consequently, adolescents carry with them a sense of who they are and what makes them different from everyone else.
Although adolescent self-understanding becomes more introspective, it is not completely introspective; rather, self-understanding is a social-cognitive construction. The developing cognitive capacities of adolescents interact with their sociocultural experiences to influence self-understanding. For example, an adolescent’s self-understanding is based, in part, on the various roles and membership categories that define who adolescents are. Stephanie is a fourteen-year-old girl who understands that she is a student, family member, hockey player, and model. Although she is at risk for HD she does not include this in her self-description; she does not regard her at-risk status as pertinent to her self-conception.

The stories of the adolescents as told by recounting the events marked on the lifelines demonstrated changes in the way the adolescents viewed themselves and their capacity to function. The theme of emerging self-understanding was illuminated by the shifts that occurred in the way they began to think and characterize themselves – that is, in their self-conceptions. The adolescents were more likely to employ complex, abstract, and psychological self-characterizations. As they began to talk more abstractly about their individual conceptions of self, they began to be able to see themselves in psychological terms. They became more descriptive in their understanding of their own personalities and discussed why they believed they behaved the way they did. The adolescents’ understanding of themselves appeared to be gradually constructed over time and represented a complex interaction between developing cognitive capacities and socialization experiences. The level of cognitive development and the adolescents’ relationships with parents, peers and other significant people in their life contributed to the development of a self-understanding.

“Strict” phenomenology instructs the researcher to listen respectfully “to what the phenomenon speaks of itself” and refuses “to tell the phenomenon what it is” (Colaizzi, 1978:52). However, as discussed in Chapter Three the problem is that we often know too much about the phenomenon we are investigating and our assumptions “predispose us to interpret the
nature of the phenomenon before we have even come to grips with the significance of the phenomenological question” (Van Manen, 1990:46). In order to obtain insightful accounts of participants’ lived experience, some phenomenological researchers make their assumptions explicit prior to doing their research. That is, they attempt to bracket- or put out of play-everything they “know” about the specific human experience they are investigating in order to prevent their assumptions from creeping back into their reflections and questions while they are doing their interviews (Van Manen, 1990:47).

I do not believe that my personal biases (assumptions) on the nature and process of self-identity and self-understanding significantly influenced the interviews, but the analysis of self-understanding was different from analysis of previous themes in that it became too complex to be able to successfully bracket out my experience and role as a therapist. In order to make this influence explicit, I drew on the historical connection I have with Harter’s work on identity development that has informed my practice since the late 1980’s (1988). Despite the assistance provided by Gottlieb’s model on the dual role of researcher/therapist, Harter’s work has become an inevitable part of the lens that I use in my work with adolescents. In order for me to provide an explicit and academically honest description of the phenomena under investigation, I have identified Harter’s dimensions and explicated how they shaped my understanding of the complex process of self-understanding and future possible self in the lives of the adolescents at risk for HD.

Harter’s (1990a, 1990b) ten dimensions of self-understanding are utilized to help clarify the theme of integrating the evolving self-understanding and the possibility of future self presented by HD. The ten dimensions are: abstract and idealistic, differentiated, contradictions within the self, fluctuating self, real and ideal, true and false selves, social comparison, self-conscious, self-protective, unconscious, self-integration.
Piaget called abstract and idealistic thinking the “formal operational stage.” In this stage individuals move beyond the world of actual, concrete experiences and think in abstract and more logical terms. As part of thinking more abstractly, adolescents develop images of ideal circumstances. They may think about what an ideal parent is like and compare their parents with this ideal standard. As Carla stated in her interview,

I always figured that boys look up to their dads. And I never had a chance to look up to my mom. I want to look up to my mom. And it scares me that I never had the chance to.

Tessa, however, found that her mother was close to an ideal.

We had to do [a class assignment on] someone who influenced us in the past year or something. And a bunch of people did all these famous people and stuff. And I just did my mom. I just think she’s really great and . . . for what she’s going through and everything. And that’s why I picked her.

Carla and Tessa were speaking about mothers who have had symptoms of HD for as long as they can remember. Carla experienced her mother’s HD as a barrier to establishing a relationship with her; it prevented her from having a mother she could “look up too.” Tessa, on the other hand, saw her mother’s ability to deal with HD as representing an ideal that she respected and she wanted to share that with her class.

The diversity of response may be influenced by the reported difference in their mothers’ symptomatology. Carla’s mother had difficulty controlling her emotions, and had been physically violent to Carla and her father, and to the caregivers in the extended care facility where her mother lived. In the beginning of my interview with Carla she stated,

I don’t remember specific events. All I remember is my mom yelling at my dad a lot and my dad telling me there was something wrong. As I got older all I remember is more violence around the house.
Not only did Carla reflect on a life full of potential and actual violence, she also knew that her home life was different from her friends. "I always knew that other parents didn’t fight like my mom and dad." Carla’s life was marked by the uncertainty of when and how violence and fighting would erupt. Her concrete experiences over time led her to believe that she would never have the mother that other people her age experienced. She saw HD as the barrier to accessing her mother.

I mean, I never really knew my mom, which is quite sad. My dad always talks about her, how she used to dance, and how she used to be so good. I would give anything just to see that. I think back about how I don’t know my mom and there’s nothing I can do about it. And today . . . I see a disease. I see somebody I don’t know but who I’ve lived with my whole life.

Tessa’s mother did not have mood swings that she remembers. In fact, she recounted that she has never known her mother to be any different than she is today, with the exception of a decrease in her mother’s ability to care for herself. Tessa’s relationship with her mother is predictable and does not include the violent reactions that plagued Carla’s relationship with her mother. But each girl knew that their friends had mothers that were more accessible and available to them. As Tessa says,

My friends get to spend more time with their moms. I’d like to do things with her and stuff. But I can’t really because she can’t really go anywhere and she doesn’t talk a lot.

The ideal mother was absent for both of them. For Carla it represented a terrible loss. Tessa considered the situation a loss, but one that gave her the opportunity to view her mother in idealist terms because of how her mother managed her disease.

The diversity of response may also be further influenced by differences in their families. Carla has no siblings and feels “alone” in dealing with the impact of her mother’s disease.

Carla’s mother is in an extended care facility. Her father is engaged to marry another woman and is involved in establishing a new life. Carla is conflicted about her future stepmother but realizes
that she will have to “live with it.” Carla feels that she does not want to tell her father about her
difficulties because she doesn’t want to worry him. “I mean he’s got enough on his mind. We’ve both got pretty hard lives.”

Tessa is the youngest of four sisters (the eldest sister was not in the study). All the girls assist in caring for their mother, although homemakers provide the primary care. Tessa’s father is home in the evenings and weekends. He participates in caring for Tessa’s mother and is active in the HD Society. He is also involved in establishing a new relationship with another woman. Tessa’s response to the relationship was similar to Carla’s. “There is something going on. If I could help it, I would. I really can’t do anything about it.” What both the girls do have in common is the realization that they cannot change the new relationships that their fathers are bringing into their lives.

The ability to think abstractly also provides the adolescent with the opportunity to entertain possibilities for the future. Adolescents are generally fascinated with what they can be. The adolescents in this research were no different, as Margaret illustrates when talking about her future self.

In a way, I can’t wait to move out because I’ve done so many things on my own. I think, hey, this is fun. I can’t wait to do it in my own home, and so I really can’t wait to move out. I know how to do so many things... like around the house, and I just can’t wait until one day I can be a mom and stuff like that.

For Margaret, the future holds positive possibilities even though she herself is at risk for HD. She perceives that the skills she learned from helping her mother cope with HD can assist her with her future goals.

In solving problems, formal operational thinkers are more systematic, developing hypotheses about why something is happening the way it is and then testing these hypotheses in a deductive fashion. Marilyn demonstrates this with the following:
[My dad] doesn’t get depressed a lot, and I always thought, what would make me depressed? And when my mom said he was going through a really hard time [I thought,] he’s going through a hard time? What is he going through? And I was kind of trying to picture it. Was it work? No, it was not work. I highly doubt it is work. So, I don’t know why I guessed it, but I just guessed. And then I thought, that’s what it is! That’s it! . . . I know why he’s depressed! He said, no, you don’t. And I said, yes, I do, and blah, blah, blah. I thought I was so smart.

Marilyn’s ability to deduce that her father’s change in mood might be caused by the knowledge that he has the gene for HD demonstrates how adolescents in a family can discover family secrets through adult behaviour without verbal acknowledgement.

The ability to employ abstract and idealistic thinking allows the adolescents to engage in cognitive behaviour that takes the concrete experiences in their lives as a basis for abstract thought. In the lives of at-risk adolescents, the development of images of the ideal parent can be diverse, and, as demonstrated in this research, may be largely dependent on the symptoms of the parent as well as the support and communication provided within and outside the family system.

The ability for abstract thought also creates an opportunity for the adolescents to consider the future. Adolescents at risk considered futures that are not dissimilar to adolescents who are not at risk. A more detailed analysis of future self will be explored later in this chapter but, generally, as Margaret illustrates, the adolescents held a positive view of their future selves.

An important part of formal operational thinking skills is the ability to develop a hypothesis and to engage in deductive thinking. As discussed in Chapter IV, the adolescents knew that “something was wrong.” The ability to use the concrete experiences in their lives to deduce a problem or uncertainty in the family was demonstrated by Marilyn’s example.

Differentiated

Adolescent self-understanding becomes increasingly differentiated. In other words, adolescents are more likely than children to describe the self with contextual or situational
variations. For example, fifteen-year-old Margaret describes herself with one set of characteristics in her relationship with her family (helpful, responsible), and another set of characteristics in her relationship with peers and friends (quiet, private). Yet another set of characteristics appears in her description of her romantic relationship (open, vulnerable). She talks with her boyfriend, and knows that he will be able to recognize when she is upset and insist on knowing how she is feeling. She appreciates his insistence, and tells him how she is feeling because “I want him to know.”

In Margaret’s case, much of her home life is private and a part of herself that she does not share with others, including her sisters. Margaret does, however, differentiate between topics she will communicate about and people she will communicate to. She will talk to her younger sister about the practical aspects of taking care of her mother. (She doesn’t communicate with her older sisters in any significant way.) She will speak about her feelings with her boyfriend, but he must press her to disclose her feelings. It may be that his persistent desire to understand how she is feeling convinces her that he really does want to know and help her deal with her confusion. She does not want to share her feelings with her friends. In the interview Margaret had a difficult time discussing her mother’s illness. She was concerned about her mother’s feelings, and did not want to disclose any information that could be construed as negative. Margaret’s ability to decide who she will speak to and what she will speak about exemplifies how adolescents are more likely than children to understand that one possesses different selves, depending on one’s role or particular context.

This ability to describe the self with contextual or situational variations was demonstrated by the seven adolescents who chose to withhold information about their personal struggles and concerns about HD from their family or medical health professionals. As described in Chapter V, the ability to delineate who to speak to and what to speak about with regard to HD is a relatively well thought out process that most of the adolescents engaged in; however, many kept their
difficulties to themselves for fear it would burden or distress their parents. It is important to realize that adolescents may appear to be handling the impact of Huntington’s Disease when, in fact, they are not managing well.

Contradictions Within the Self

The differentiation of the adolescent self into multiple roles in different relational contexts naturally leads to potential contradictions between these differentiated selves. Emma at one point in the interview states, “I’m strong. . . . I hold everything in.” Later on in the interview Emma offers a contradictory self-description: “I’m kind of one of those people that anything you say . . . I’ll cry very easily. It doesn’t take much.”

Adolescents develop the cognitive ability to detect these inconsistencies, as Daphne illustrates when she ponders whether she should have children in the future, given that she may carry the gene for Huntington’s Disease.

Part of me is . . . I want children! And the other part of me is like, well, you know, you should do this and blah, blah, blah. My conscience kicks in and I don’t know if I could deal with that either. I want to do something, and then I don’t know what I want to do with my life. It makes me think about things, and I actually have to plan, which is good for me because I don’t plan. [Laughs] It makes you feel philosophical in a way. It does, you know, like actually having to plan and do some thinking when I am a spontaneous person.

Although in other parts of the interview Daphne talked about her fear of having the gene for HD, when using the construct of creating a general theory of self, Daphne can be seen as an adolescent looking toward the future in a way similar to that of any teenager. The at-risk

34 In one study, Susan Harter (1986) asked seventh-, ninth-, and eleventh-graders to describe themselves. She found that the number of contradictory terms used to describe oneself (moody and understanding, ugly and attractive, bored and inquisitive, caring and uncaring, introverted and fun-loving, and so on) dramatically increased between the seventh and ninth grades. The contradictory self-descriptions declined in the eleventh grade but still were higher than in the seventh grade.
adolescents spoke about contradictions in their behaviour as any adolescent would. Huntington's Disease did not seem to significantly influence the differentiating of self.

The Fluctuating Self

Given the contradictory nature of the self in adolescence, it is not surprising that the self fluctuates across situations and across time. Daphne illustrates this when speaking about how she has been feeling about her life: "I have been pretty happy over the last little while. I mean I have had a couple of ups and downs, because, you know, I am still pretty depressed lately." The adolescent's self continues to be characterized by instability until the adolescent constructs a more unified conception of self, usually not until late adolescence or even early adulthood.

Korer and Fitzsimmons (1985:589) found that parents would often monitor their at-risk offspring's physical and emotional behaviour, as demonstrated in this quote from a parent in their study.

I worry nights since I found out it could be passed on. I had terrible nightmares. I keep looking for them doing things they wouldn't normally do, and then I can't do anything about it.

It may well be that parents' lack of understanding about the normal dimensions of self-understanding that evolve in adolescence distorts their view of their child. Reactions to the adolescent's behaviour may be affected by the parents' apprehensions surrounding HD, and misinterpreted as a symptom of the disease rather than a natural part of adolescent development.
Real and Ideal, True and False Selves

The adolescent’s emerging ability to construct an ideal self leads to the capacity to differentiate between real and ideal self. While this represents a cognitive advance, it can also create confusion for the adolescent.35

As discussed in the literature review, an important aspect of the ideal or imagined self is the “possible self” (Markus & Nurius, 1986). The possible self is what individuals might become, what they would like to become, and what they are afraid of becoming.

Kathy’s comments demonstrate how the future positive self can influence future directions.

Yeah, I’d like to do that [work on a cruise ship] ‘cause you make pretty good money and I could pay my student loan off. And I think if this hair thing doesn’t work out ... I would take a course to be a homemaker. My friend took the course ... it was funny, she asked me last week to go to her graduation supper, and they were just kind of talking a little bit about the course and stuff. She knew I was interested in that and that we’d had homemakers in and out of our home. So, it’s kind of neat.

Carla’s comments reflect on the possible negative self that HD presents.

I look at my future as obviously me becoming a singer. But I also see another path that might happen. And I watch my mom, and I see me in the future and that scares me. My friends always tell me [that I’m not going] to get it. ... But I’m always looking to the other side. I can’t help it.

Carla’s reflections on the possible negative self that HD presents did not become a conscious consideration for her until she turned fifteen. For adolescents at risk for HD, this could be the time when their at-risk status becomes challenging, as Carla’s comments indicate.

I don’t think I ever really thought about it until I was about this age. I was like, all right, I have a 50% chance of getting it but I don’t care about that right now; my mom is what I’m focusing on. I still have [some time before] it could actually kick in, but this is now, and my mom’s got [HD]. I never really cared about myself.

35 Researchers have found that the discrepancy between the real self and the ideal self is greater in middle adolescence than in early or late adolescence (Strachen & Jones, 1982).
Marilyn found that she began to consider her risk status and the possible negative implications on her future self when, at sixteen, her family members started asking her questions about whether she would want to be tested.

I look into my future because I have to right now. I’m in grade twelve and my whole life is in front of me. My mom and my aunts ask me if I’ll want to know if I have the gene when I’m eighteen. Are you going to get tested? Are you going to find out? It is their questions that made me start to think about my possible future. I don’t know, I might [get tested], just to know, but I don’t think it would change me.

The ability to distinguish between real and ideal selves is a dimension in self-understanding that occurs in adolescence. The aspect of the ideal self that Markus and Nurius (1986) explored can be helpful when considering how at-risk adolescents consider what they might become or what they might be afraid of becoming.

In this study, seven adolescents indicated that they did not actively think about their future selves in negative terms (that is, in terms of HD) until they were fifteen or older. Marilyn was contemplating what she would pursue upon graduating; it was when questioned by her mother and relatives about predictive testing that she shifted her awareness to what she was afraid of becoming.

Tessa and Stephanie, who are both under fifteen years of age, did not speak about a fear of having the gene for HD. Stephanie felt that a cure would be found in “two years,” and Tessa “[doesn’t] think about it a lot.

**Social Comparison**

Adolescents are more likely than children to use social comparison to evaluate themselves. Marilyn demonstrates this evaluative function of social comparison.
I find people fascinating, and in a way it just helps me with my life if I observe other people’s lives. I don’t necessarily observe the bad things because, personally, I observe what they do well so I can do it better. Competitive, you know! [Laughs] Someone hits the ball up and I think, I can do better than that, and I hit it farther.

Relying on social comparison information in adolescence may be confusing because of the large number of reference groups. For example, should adolescents compare themselves to classmates in general, to friends, to their own gender, to popular adolescents, to good-looking adolescents, to athletic adolescents? Simultaneously considering all of these social comparison groups can be overwhelming for adolescents.

Margaret reflects,

I was a junior counsellor at a camp [with kids] similar to my age, so the age between the campers and me wasn’t that big. And some of the boys – you know how boys that age are – they think they’re all tough. And they’d just put on this big act sometimes, and they’d try to tell me that I wasn’t smart or things like that. They just walked all over me. And I couldn’t understand why because I was sticking up for myself. I couldn’t understand why it was happening to me and not other junior counsellors my age, like girls. And I still don’t understand why that is.

Given that the adolescents in this study were much like any other adolescents developing the skills of comparison to assist them in their self understanding, Marilyn shows how this dimension of social comparison was affected by being in a family with HD. She spoke of a time when she was thirteen years old and understood about the hereditary nature of HD. She began to compare herself to her friends. It was at this time in her life that she began to have “a lot of self pity.”

I’d just think, why is my mom sick? Why does my family have HD? Why do I have to go through this, you know? ‘Cause all my friends, their moms weren’t sick. And I was always wondering, why aren’t their moms sick, you know? And what really makes me mad is when one of my friends would get mad at their mom. I’d be like, you don’t know how long you’re mother is going to be there and your mother is one of the best people in your whole life – she gave birth to you; how can you talk to her like that?
Five of the adolescents in this study spoke of having the experience of comparing themselves to others outside their family who were also at risk for HD or who had the gene for HD. Kathy, Margaret, Tessa, and Emma attended a national Huntington’s Society conference where I facilitated an at-risk adolescent support group. Margaret, Tessa, and Emma attended the adolescent group while Kathy, who was nineteen, attended a support group for adults at risk for HD. Kathy found that the adult support group was helpful, and that many of her feelings about being at risk were shared by others.

I went to a meeting for at-risk people. And a lot of them were hesitant about getting tested for HD. I think there were two girls that were in the middle of getting testing. The rest of us were, you know, hesitant. Even forty-year-olds were hesitant about getting tested. It was helpful to sit with a group of people like me – how often does that happen? Just to hear about how people cope. The man who led the group would ask us questions. It was a good discussion, and we could compare our ideas with each other.

The adolescent support group was for teenagers between the ages of thirteen and nineteen. They were told the session would be private and confidential and that I would be the only adult in attendance. A medical geneticist came into the meeting and insisted on staying. The adolescents were noticeably uncomfortable, and I asked them if we could use the first part of the meeting to clarify any medical questions they might have as we had a geneticist to answer questions. She answered many questions, and after thirty minutes I thanked her and suggested that she attend some other group where her assistance might be required. After she left, the adolescents talked about how they had thought they were the only ones who didn’t understand about probability of risk; they expressed that they felt better knowing that other adolescents at risk also experienced misunderstandings around HD. The group went on to discuss other more personal topics, such as dealing with affected parents and talking to friends. Margaret, Tessa, and Emma told me in each of their interviews that it was the first time that they could compare themselves to other adolescents who were at risk, and that it made them feel better about how
they reacted. As Tessa recounted, “It was the first time that I realized that other kids had a tough
time, too, and how lucky I am that my mom is so nice.”

As mentioned earlier, Daphne attended a monthly Huntington Disease Society meeting in
her city. She compared herself to people who were symptomatic, and saw her own “mortality.”
She found the comparison very uncomfortable and chose not to attend another meeting, as it was
too difficult for her to see her possible future.

The adolescents in this study were much like any other adolescents utilizing social
comparison as a way to help increase self-understanding. These skills of comparison were used
by five of the adolescents when in groups that consisted of at-risk people or people who had the
gene for HD. Four of the adolescents found the experience useful; in particular they felt that the
experience normalized their own responses to HD and to the affected parent. All four felt a sense
of relief that they were not alone in the feelings or behaviours that they experienced living in a
family with HD. Daphne was the only adolescent who felt that the experience was too difficult,
as it brought to mind her feared future self.

Self-Conscious

Adolescents are more likely than children to be self-conscious about and preoccupied
with their self-understanding. As part of their self-conscious and pre-occupied self-exploration,
adolescents become more introspective. However, the introspection is not always done in social
isolation; sometimes adolescents turn to their friends for support and self-clarification, obtaining
their friends’ opinions of an emerging self-definition.

Daphne describes this when she speaks of her younger friends.

They remind me of me when I was that age. I really identify well with them
because they know part of what I have gone through and they have been through
similar incidences . . . and so we have a basic comparison and something to talk
about. And some of these young kids are amazing. I am just totally stunned by the
intelligence of some of these kids.
Adolescent friends are often the main source of reflected self-appraisals, becoming the social mirror into which the adolescent anxiously stares. As discussed in the literature review, this self-conscious, self-preoccupation reflects adolescent egocentrism. The adolescents in this research reflected this egocentrism in the form of two types of social thinking: imaginary audience and personal fable. The imaginary audience refers to the heightened self-consciousness of adolescents that is reflected in their belief that others are as interested in them as they are in themselves.

Tessa demonstrates the effect of this belief when talking about lack of transportation in her family.

Since my mom can’t drive, and I still can’t drive, and my dad is often gone at meetings at night or things like that, I am often left with no drive home and I have to walk and people usually say I’ll give you a drive. Sometimes I know that people are feeling bad for me. You can just sense from the way they’re looking and talking at me. They think, Oh, you poor girl, your parents... I feel like they’re looking down at me, as if my parents don’t care. But I just don’t really want to... explain to them why. Like, I know that’s the only way I can get rid of it, but, like, my boyfriend’s parents, they drive me home an awful lot when I’m over. And they come and pick him up when he’s at my house and they take us places. And the odd time my dad or my sister will come to get me. I think they understand why because they know my dad and they’ve met him before. And Tina’s parents understand, too. But there are not many other people who do. I don’t like being judged or looked at.

The personal fable is the part of adolescent egocentrism involving an adolescent’s sense of uniqueness. As part of their effort to retain a sense of personal uniqueness, adolescents may craft a story about the self that is filled with fantasy, immersing him or her in a world that is far removed from reality. Carla demonstrates this in her perceived deep connection with a rock group called the “Spice Girls.” She mentioned them frequently in her interview, and spoke of them as if they were close friends who understood her unique position in life. One member of the
Spice Girls named Melsie became a sort of substitute for Carla's mother; a mother figure that she could "look up to."

I mean, I never thought you could love someone so much without knowing her. But when I look at Melsie it's so weird. I would look up to her more than I would look up to my mom. I see all these families... they're so happy. I come home [and wonder,] why couldn't I have that? I mean, I think that's why I look up to the Spice Girls so much. Because if you look at them, their friendship is so much stronger than so many other friendships, and I look at my and Sophia's and Clara's friendship and there's something missing. Seems like everything I have there's something missing.

Through a fantasy relationship with a music group, Carla was able to establish a place for herself within a family of sorts. Her mother had been a dancer, and Carla's father told her stories of how well her mother danced. Carla dreamed of being a famous singer, and wished her mother could see her perform. As noted earlier, she longed to have seen her mother's artistic expression, and she longed for her mother to witness her own artistic endeavours. It was within the realm of music – a possible crossroads of worlds where she and her mother could have met – that she found the Spice Girls. They became her fantasy family, where she could feel like she imagined her friends felt, and where she could create a parent-like figure that she could look up to.

The adolescents in this study were much like any other adolescent when examining self-consciousness and introspection. The two types of adolescent egocentrism – imaginary audience and personal fable – were demonstrated as being part of how the adolescents dealt with the impact of HD in their lives. Carla's poignant example demonstrates how adolescents cope with living with a parent whose symptomatology creates a barrier to a normal mother-child relationship. The ability to create a fantasy world can be viewed as a positive coping strategy that may assist Carla in developing a healthy sense of self-understanding despite the loss of connection to mother and family that she believes is a result of living in a family with HD.
Self-Protective

Although adolescents often display a sense of confusion and conflict stimulated by introspective efforts to understand the self, they also call on mechanisms to protect and enhance the self. In protecting the self, adolescents are prone to denying their negative characteristics.

John talks about dealing with his depression in a way that reflects this self-protective mechanism.

So, I started in grade eight and I made some friends – fair day friends. . . . But I made some friends with my sister’s friends. I started hanging out with them a little bit more, but I was still a pretty depressed kid. And then we moved and I decided that I wanted to change myself. So, I cut off all my hair. I cut all my hair. I changed my name from Johnny to John – dropped the “y.” And I moved to my new school.

Although the adolescents in this research are like any other adolescents engaged in developing self-understanding, it is informative to learn how these aspects of identity formation are utilized when dealing with the impact of Huntington’s Disease in their lives. As stated earlier, the adolescents did not begin to consciously consider the negative aspects of their future until they were at least fifteen years of age. The seven adolescents in this study who were over fifteen years of age disclosed to me that they were talking about their possible future for the first time. Emma used a positive self-description when she considered how she would imagine her self to be if she were to have the gene for HD, “And, that’s what I’d be like, a nice person with Huntington’s.” The other six adolescents who were over the age of fifteen did not use self-descriptions when talking of their future self should they have the gene for HD. They spoke instead about what they would do or not do if they had the gene. They talked about how they would live their lives more fully or “compact.” It may be that the complicated process of understanding the future implications of being at risk in terms of core aspects of self require more complex cognitive functioning than young adolescents are capable of. However, as Emma
illustrates, some adolescents are able to consider their future self while still protecting and enhancing self-identity.

**Unconscious**

Adolescent self-understanding involves a recognition that the self includes unconscious as well as conscious components, a recognition not likely to occur until late adolescence (Selman, 1980). Older adolescents are more likely than younger adolescents to believe that certain aspects of their mental experience are beyond their awareness or control. Nineteen-year-old Kathy speaks about her awareness of being at risk for HD.

> I know it's there. It's always kind of in the back of my mind, just like the rest of all that kind of stuff. But I really learned to take each day at a time because you just get yourself in more of a mess if you don't.

As discussed in the dimension of “self-protectiveness,” the adolescents in this research perceived being at risk differently at different developmental stages. As illustrated by Kathy’s comments, it was a more predominant concern for older adolescents. The younger adolescents in the research were more concerned with the concrete experiences of their everyday lives. As Stephanie explains, “I know I’ll just be older if it happens [become symptomatic] and it won’t bother me.” She believes that there will be a cure; her focus is on how to deal with the impact of her father’s behaviour in her life. She, as well as Tessa, did not discuss any awareness about the unconscious influence that being in a family with HD or being at risk for HD might have in their lives.

Exploration of the adolescents’ at-risk status will be more fully developed in the section on future self.
**Self-Integration**

During adolescence self-understanding becomes more integrated, with the disparate parts of the self more systematically pieced together; this is particularly true of late adolescence. Sixteen-year-old Marilyn displays this task of integrating when discussing her future self.

My whole goal is to find a job that I love to do. It doesn’t matter [about] the money. You ask people now at this age, what do you want? Oh, I want to be rich. It means nothing to me . . . I guess because I’ve grown up with it. Like if you look at my mom; I guess this is the reason why I don’t want to be rich. She grew up in a really poor place. And her whole dream was to marry a lawyer and become rich. And that is what she got. She got that. She married a lawyer. She is rich and she is not happy at all. It doesn’t bring anything. And I guess because I grew up with living in a nice house all the time and nice cars I don’t see any comfort in that. I don’t want to be poor and on the streets. But I do want just enough to be happy. And I guess what I want is to find a job that makes me happy.

It is interesting to note that Marilyn compared herself to her mother, the parent that does not have HD. This may be due to the fact that Marilyn’s mother provided a consistent and stable role model in her life. She described her father as seldom being available for her when she was growing up, and how, with the onset of his symptoms, his behaviour became erratic and unpredictable. It may be that it is his absence in her life when she was growing up rather than the onset of symptoms of HD that has caused Marilyn to focus more on her mother in shaping her general concept of her self and her values. Marilyn demonstrates how adolescents attempt to integrate various parts of their understanding about self to come up with a more cohesive sense of self.

As stated earlier, self-understanding is a social-cognitive construction. An adolescent’s developing cognitive capacities interact with socio-cultural experiences to influence self-understanding. The narratives of the adolescents in this study illustrate these dimensions and the complexities of self-understanding as reflected in the exploration of the ten dimensions of self-understanding. While the cognitive component of adolescent development is clearly unaffected,
the socio-cultural component is affected by the inclusion of family experiences of HD, reactions of others, and the growing recognition of personal risk.

Future Self

Planning for the future is not a simple task for any adolescent or young adult. Decisions around choosing a career, deciding about post-secondary education, and finding a mate are just a few of the challenges that adolescents face. When this is coupled with knowing that one is at 50% risk to get HD in adulthood, it can make viewing the future and choosing future trajectories a difficult task. This section of the chapter will examine the adolescents’ view of their future, personal meaning of at-risk status, and their views on presymptomatic testing.

Views of the Future

Most research into HD has not explored the at-risk adolescent’s view of the future or the impact of making decisions about predictive testing. Not only is there a lack of research in these areas, there is a fear of the emotional reactions that might be generated by a discussion of this nature (Korer & Fitzsimmons, 1987).

I certainly felt that this research put me in unknown territory. I did not know what to expect from the adolescents. It was the case that for all the adolescents in the study I was the first person to ask them questions about the future. The first pattern to emerge was the adolescents’ existential sentiments about the meaning of life and relationships that would not be developmentally expected for their age range. In fact, a number of the adolescents reported feeling older than their age owing to living in a family with HD as well other challenges in their lives. As Emma declared, “I have been through a lot in my life; I am really like thirty-six-years old. [giggles]” Kathy believes that she is “old before her time” because of all the challenges she has faced in her life, including having a symptomatic mother. Daphne also felt older than her
age: "Most of my friends are like, you were twenty-one when you were sixteen. I have had a lot of experiences and I identify better with older people."

Having a parent with HD caused each of the adolescents to think about their future in what appeared to be more deliberate ways; they simply did not take things for granted as many young people do. Each adolescent had their own unique approach, shaped by an awareness of their parent’s physical and mental deterioration and the possibility that they might also have the gene for HD and thus a shortened life span. There was, however, a common theme of hope for the future. Most of the adolescents held a belief that they could create a meaningful life whether they had the gene for HD or not. Four themes of the future self emerged for the adolescents: quality not quantity; staying in the present; future generations, it’s not the HD.

Quality not Quantity

Four of the adolescents felt that they would live a life that was so full of experiences that it would not matter if they had the gene. For instance, Stephanie believes that she will just live her life "faster." "I will probably do more than most people, and do it a lot faster." Carla believes that she will plan out her life into a "kind of compact space; I want to do as much as I can." As Marilyn states,

I want to do everything that I can. If I have it, I want to do as much as I can because I don’t want to live my life to the end if it’s going to be so . . . I don’t know. I mean it probably won’t be the way I picture it. It probably won’t, but because of growing up seeing it that way, I just don’t want it.

For John, being at risk means that he may have to "condense" or "rush" his life. When he speaks of his future dream of working in China he says, "You know, I would stay a year in China instead of staying five years in China if I had my whole life to live without HD. With HD I might make it a year and move on to somewhere else to see other things."
Staying in the Present

Three sisters, Tessa, Margaret and Kathy, all had the philosophy of dealing with their future risk of HD when it happens. Tessa, the youngest sister, states, “I don’t think about it a lot. I want to be a dentist, and if I have HD I will shake too much so I don’t think about it.” Tessa plans to live her life with the goal of becoming a dentist and worry about HD when she is “much older.” Margaret, the next oldest, has similar feelings.

I always have known that I want to do something in business, but I never thought of what happens if I do have Huntington’s? What am I going to do about my career? And the way I see it now is that I’ll think about it when the time comes. Cause this is what I want to do now, and if that happens, if I don’t have it, great, then I’ll continue on, but I’ll see what happens when the time comes.

Kathy, the eldest of the three sisters interviewed, believes that having the possibility of HD in the future “probably has impact.” She feels that HD is “far away.” She believes that she is “prepared now for HD if it ever hit me” but wants to live in the present for now. When I asked her how she is prepared, she replied, “I’ve seen my mom handle it and that has prepared me.”

Future Generations

Daphne’s dream of the future is to have children. The idea of HD as a risk presents the challenge of how to decide whether to adopt children or to have her own, knowing that they may be at risk for HD. Her grandmother is a confidant, and has spoken to her about the challenges of raising a son (Daphne’s father) who has HD. This causes Daphne concern, and leads her to wonder whether it is “fair to the child to be put in that kind of position.” Interestingly, Daphne did not reflect on what it was like for her to be in the very position that concerns her when she thinks of her potential children. And Daphne never addressed the fact that if her children have the gene for HD she would have the gene for HD and might also be symptomatic.
For Emma, it is not Huntington’s Disease that looms as a problem for her when she considers her future.

It’s not the Huntington’s that bothers me. It’s just my dad’s temper that bothers me. If he were a nice person with Huntington’s, it wouldn’t bother me, I wouldn’t care. And that’s what I’d be like, a nice person with HD. But, he is not. HD – that doesn’t bother me at all. I don’t care. I get it, I get it. I don’t care as long as I live a long time.

Emma was the only adolescent in the research who had a parent with another disease. Emma’s mother had cancer, and Emma stayed home to be with her mother as she was dying. Emma was very close to her mother and felt included in her life in a safe and intimate way. Emma revealed that in the past her father had physically and verbally abused her. She believed that her father had this problem before HD and could choose to be kinder to her if he wanted to. Emma seemed to feel that everyone has a choice to conduct themselves appropriately with family and non-family members, whether they have HD or not. She reported that the most important factor in her life was her father’s anger and the fear and uncertainty it created in her life. Whether she looked at the future or the present, her main concern was to exist in an atmosphere of safety. Huntington’s Disease was an issue in her life, but only one issue among many other concerns.

When discussing their future selves, most of the adolescents believed that they could create a meaningful life regardless of their gene status.

At-Risk Status and Predictive Testing

As stated above, the adolescents carried with them a common theme of hope for their future. It manifested in the way they looked at their future goals. This theme of hope for the future was also expressed in their belief that the gene discovery and current promising research
would lead to a cure and thus change the fateful destiny of their families. A variety of perspectives emerged as the adolescents began to describe how they experienced their at-risk status. When discussing at-risk status, seven adolescents included the topic of predictive testing and whether or not they were interested in taking the test. Two of the adolescents did not discuss predictive testing.

The adolescents’ perspective on their at-risk status will be discussed using four themes that emerged from talking about predictive testing: desire for future knowledge, fear of future knowledge, uncertainty about future knowledge, indifference about future knowledge.

Desire for Future Knowledge

Three adolescents (Carla, Emma, John) were clear that they wanted to take the predictive test for Huntington’s Disease. Carla was ten when she was first told that her mother had HD and that she was at 50% risk for HD. She was attending the HD clinic where her mother had received predictive testing. When I asked her what information she walked away with from her session, she replied, “I walked away thinking that my mom was dying. And I didn’t care that I had it, that I had a chance. I was just like, no. My mom can’t leave me.”

It wasn’t until Carla, at fourteen, took a tour with the Huntington Disease Society of BC through the Centre for Molecular Medicine and Therapeutics research lab that she began to understand what it meant to be “at risk” “I learned a lot about my risk status on the tour, and that’s when I think I became the most worried about the fact that I have a 50:50 chance now.” As stated earlier, Carla is “really, really scared” because she sees the possibility of two paths before her.

I look at my future as obviously me becoming a singer. It may become something like having a huge career. But I also see another path that might happen. And I watch my mom, and I see me in the future and that scares me. A lot. I mean it’s so hard to explain. I don’t think anyone can really understand.
Carla went on to explain that there is not only the fear of becoming like her mother but also the reality that there have been many secrets in her life, and she would like to have some control over her own life.

Taking the test would take some of the unknown away and let me plan my life. All of my life I’ve pretty much been unknown to everything. I never really fully understood anything, and people kept secrets from me. So, I’d like to know at least one thing. I mean that’s one of the most important things to me right now, besides the fact that my mom has it. I mean that’s the most important thing to me, but I’d also like to know if I have it.

Carla laments, “They just don’t think a fourteen-year-old could handle it. I mean that’s not fair. I do know the decision I’m making, that’s why I want to take the test.” When I asked her if she will take it when she turns eighteen she responded, “Yeah, but I just want to have it now, by then I won’t have changed my mind. At least I’ll be able to know what’s going to happen in my future.”

Emma is not as certain as Carla about how she finally determined that HD was hereditary and that she was at risk for HD. “I think I figured it out on my own, somehow. I probably heard it from family or somewhere. I find things out, and I don’t know where they come from.” Emma emphasized to me that she wanted to “get tested NOW, but nobody will do it!” When I explored with her why people were opposed to her getting tested she explained,

Sharon [HD social worker] thinks I have a little too much stress to be thinking about getting tested. But in the papers I found about HD it says you can get tested from seventeen years of age and up. When I asked Sharon and all the people, they said, no, don’t do it now, don’t do it now. I guess they’re afraid that if I find out I have it that it’ll be just one more thing that I have to deal with.

Emma is planning to move away from home to the city and “get a job and maybe some schooling.” Once she is settled in town and has more direction in her life she plans to take the test so that she can “plan out my life.”
John was eighteen years old when I interviewed him, and he had already decided to take the predictive test for HD and had set up the date for the testing, which was to follow six months after the interview. He had learned about his risk status when his father asked the family to attend a meeting with a mental health professional. He, like Carla, walked away from the meeting hearing that his father was going to die and that he was at-risk for the same disease. John talks about the process he went through around his decision to get tested.

I pretty much started the process of thinking about predictive testing at sixteen, right after the meeting with the family about HD. I thought about it right away. I think my initial reaction was, no. It was like if I don’t know, then I can ignore it and go on with my life. And then I came to the realization that it was probably more detrimental to me not to take the test. I am pessimistic, and I will always be thinking that I have HD.

John is studying computer technology and has searched the Internet for information about HD. He believes that prevention is something he can learn about, and is another reason to have predictive testing.

It would be unfair to me not to take the test. To say to myself, I don’t know why it is happening if there is someway that I can help to put it down the road a bit – maybe put it down ten years, because I could do all this preventative stuff. Then I’d have ten more years doing more things I have to do. [It’s better] than just forgetting about it and missing those ten years.

I asked John if he has read much on the Internet about prevention, as I had not found any information about preventing HD. He admitted that he “didn’t find much,” but stated that he believes that there is “going to be a computer program that he could use in the future.”

John discussed his decision to have predictive testing with both his father and his mother. His father, who is symptomatic, didn’t think he should get tested. John said that his father thinks “it is better if you don’t know ‘cause then you will find things a lot easier.” He said that his father “just got quiet” and would not discuss it further with John. John says that his mother thinks that it’s the “right decision for me, and that I can handle it.” Both of John’s parents are
coming to his first predictive test meeting to ensure that he is “stable enough to take it, to do it.” John has also thought about who he would share his test results with. He is certain that he will tell his father, mother, and sister. “If I don’t have it then I am telling the world! But if I do have it I’ll probably tell everyone. I probably will. All the people who need to know.”

Three adolescents want to take predictive testing. For Carla and John it is the fear of having HD that makes them want to take the test. John also believes that if he has the gene for HD then the knowledge of his status may lead him to uncover a preventative treatment. Carla also wants to take some control over her life, which she feels has been surrounded by secrets. Emma would like to take the tests so that she can plan her life. Emma says that she is not concerned about HD – “If I get it I get it” – and believes that even if she had HD she could be a “nice” person.

Fear of Future Knowledge

Two of the adolescents (Daphne and Margaret) do not want to have predictive testing. Daphne, like her brother, John, attended a family meeting with a mental health professional and learned of her father’s HD and that she was at risk for HD. I asked her what she learned from the meeting. She replied much like her brother: “It was that I have a good chance of getting it. And that my dad is going to die from it. Those were the two things that stood out in my head.” Daphne and her brother have discussed predictive testing, as Daphne recounts.

We did talk about this [PT] a lot. He said that he is going to go get tested. I think that is very cool. I mean, I am not going to, but he is more of the mindset that he would rather know now. I am not. I’d rather wait. I think that if I had something like that hanging over my head that I don’t know if I could actually live with it. I would get into a deep depression and never come out. That is not what I want to do with my life – not at all.
As I explored with Daphne why she thought she would have a deep depression if she found out she had the gene, it became clear that she believes that she would handle the information in the same way as her father.

I noticed when Dad found out [PT test results] that he didn’t leave the house. He became a total recluse, like stuck in the house. He was very embarrassed about having HD movements. You know, like what if some people saw him fall, or something like that. I would like to think that I would handle it in a different way but I don’t know.”

Margaret does not think about Huntington’s Disease. “I am not going to let something that might happen in the future affect me now. If one day I get it, then I will work something out then.” Margaret attended an adolescent-at-risk group at the National Huntington’s Society Conference that I facilitated. Margaret told me afterwards that she thought most people would feel the way she did about Huntington’s Disease and predictive testing. She was surprised to find that she felt differently than most of the group. “It was kind of weird. I mean it is hard to know what people think because we don’t even talk about it in our family. I just thought most people thought like me.”

Both Daphne and Margaret did not want to take predictive testing. Daphne discussed testing with her brother, and based her decision not to test on her father’s reaction to his test results. She feared she would fall into a depression much like her father. Margaret, on the other hand, felt that predictive testing was something she did not want to consider because she wanted to follow her career plans and worry about HD later. After attending an adolescent support group at the National HD Conference, she was surprised to find out that most of the adolescents did not have the same perspective that she did.
Uncertainty about Future Knowledge

Marilyn and Kathy were uncertain about predictive testing. Marilyn started out the conversation about her at-risk status in a poignant manner: “In some ways I think, why live if I am going to have it? If I have it then there is no point to living.” As I explored her sense of hopelessness, it became clear that her experience with her grandfather in the latter part of his life profoundly affected her view of the course of the disease.

I wouldn’t want to live my life until I’m eighty if it means forty years of doing what my grandfather did. I guess that’s why I associate the end with my grandfather because that’s what I saw.

Based on her concern about the prospects of dying a horrible death like her grandfather, Marilyn is ambivalent about predictive testing.

I don’t know if I would want to know or not. I guess in a way I feel that there might be a cure out there when I am at that stage. I don’t know. I feel like if I know, I know. If I don’t, I don’t. I am going to live my life as far as I can. Well, if I have it then there is nothing I can do. I can’t go back. I’m just praying that there might be a cure by then.

Marilyn has decided not to decide about testing until she is older, and ended this part of the interview with the hope that a cure will be found so that she does not have to make the decision.

Kathy only briefly discussed her ambivalence about predictive testing. She feels that she is “still young” and has “so much to live for.” She feels that she will face HD and decisions to take the test when she is older and “wants to have children.” She feels that the example of her mother coping with HD has taught her that she can face it if she has to. Testing is not something she wants now, and she is not sure if she will want it in the future. She attended a group session for at-risk adults at a national conference of the Huntington’s Society. She felt that her feelings around predictive testing were validated because most of the people in the group session were
“hesitant” about testing. She was surprised that “even forty-year-olds were hesitant about getting tested.”

Marilyn and Kathy were not sure if they would get tested. For Marilyn, the fear of dying like her grandfather weighed heavily on her mind, and she felt that it was probably better just to live her life and pursue her goals while “praying” for a cure. Kathy also didn’t want to focus on future risk. She has had a different experience of a family member with HD symptoms than Marilyn. She has watched her mother cope with symptoms since she was ten years old, and feels that it has prepared her if she should have the gene. She may consider predictive testing in the future if she considers having children, but even then she is still unsure whether she will engage in testing.

Indifference about Future Knowledge

Stephanie and Tessa (the youngest participants in the research) were indifferent to predictive testing for different reasons. For Stephanie, there is no doubt in her mind that there will be a cure for HD.

HD does not scare me – not at all. I know they are going to have a cure in like two years. That’s what I heard. It was supposedly on the news, and my mom said it will be in at least two years. . . . So, I’m not scared. There will be a cure when I am older, and that is so far away.

Tessa is the youngest of the three-sister sibling group I interviewed. She does not “talk to anyone about anything” as she is too involved with her concern about her father’s girlfriend and how that impacts on her life. Like Stephanie, she feels that HD is “far away,” and is more
concerned with how her own life is unfolding now. Both girls are younger adolescents, and their indifference to testing may reflect their stage of development.\textsuperscript{36}

Summary

Despite the fact that each adolescent had their own experience of being at risk for Huntington's Disease, all of the adolescents thought about their future in what appeared to be deliberate ways. The seven adolescents over the age of fifteen developed more complex ideas about their future, the possible implications of predictive testing, and the possibility of a future impacted by having the gene for HD. The two adolescents under the age of fifteen had a more difficult time imagining the future, and were, in fact, far more concerned with the present effects of living with a symptomatic parent. All of the at-risk adolescents expressed a common theme of hope for the future, and most of them held a belief that they could create a meaningful life whether they had the gene for HD or not.

When the adolescents contemplated their future possible selves, seven of them initiated conversations about their thoughts on predictive testing. Only the two youngest adolescents did not discuss predictive testing when considering their future possible selves. Of the seven adolescents who discussed predictive testing, two did not want to take the test, and five either wanted the predictive test or were still uncertain about whether to take the test.

Harter's (1998) ten dimensions of self-understanding helped elucidate the complex process of self-understanding as a part of identity formation, and aided in discerning what aspects of adolescent development may be exacerbated by the impact of living in a family with HD and at-risk for HD. Not all the dimensions of self-understanding were impacted by

\textsuperscript{36} Piaget (1954) believes that we go through four stages in the process of understanding the world. The formal operational stage appears between the ages of eleven and fifteen. In this stage individuals move beyond concrete experiences and begin to entertain possibilities for the future. The indifference reflected by these two adolescents may be owing to their being at the beginning stage of formal operational thought and cognitive development.
Huntington's Disease, but clearly the adolescents carried the additional stress of HD into the process of self-understanding. Their experiences and their views on predictive testing can be seen to contribute to their self-understanding and understanding of future self, and to reflect an individual process influenced by the cognitive, developmental, and socio-cultural context of their lives.

Adolescence is an important time in the development of self-understanding and self-identity. It is a complex time during which childhood identities begin to integrate with the emerging adolescent identity – an ongoing process of identity formation leading to adult maturity. For the participants in this study, it was also a time when factual and experiential learning regarding Huntington’s Disease was integrated with an emerging recognition of their own risk status. Moreover, the integration of this knowledge occurred while watching a parent progressively lose both physical and mental functions as a result of the disease.

The experiences of these adolescents illuminate the complex process of adolescent development and identity formation, and the fact that they are dealing with the additional burden of a parent with a degenerative disease and with the challenges of their risk status points to their incredible resiliency.
CHAPTER VIII

STRUCTURE OF THE ADOLESCENT EXPERIENCE

Summary of Purpose and Results

This research study investigated the experience of the adolescent at risk for Huntington’s Disease. Nine adolescents between the ages of thirteen and nineteen years were interviewed about their experience of living in a family with HD and being at risk for HD. The interview and the analysis of the interview content was conducted according to the tenets of phenomenological inquiry, particularly as set forth by Van Manen (1984). The analysis of the commonalities in the co-participants’ experiences was also informed by the principles of narrative research, particularly as set forth by Cochran and Claspell (1987) and Cochran (1990).

An analysis of the interview data resulted in the identification of three themes and a fundamental structure – or common story – of the co-participants’ experiences of living in a family with HD and at risk for HD. The three themes are: (1) Naming the Legacy: Understanding and Misunderstanding; (2) Experiencing the Legacy: Huntington’s Disease in Relation to Relationships; (3) Integrating the Legacy: At the Crossroads of Self and Future Self.

In order to provide an overall structure of the experience, an interpretation of the analysis in light of previous literature pertaining to adolescents at risk for HD and adolescence in general is presented.

Huntington’s Disease and the Adolescent Experience

This study illuminates the adolescent’s subjective understanding of being at risk for Huntington’s Disease and living in a family with Huntington’s Disease during an important period of identity formation. It is unique among studies focused on adolescents at risk for Huntington’s Disease due to its phenomenological methodology and the limited research in the
subject area. As a consequence, its results can be compared only in a general sense to previous research in the field, most of which emerged from quantitative studies. Moreover, this study was not theory-driven, and was not designed to test the validity of any particular theoretical perspective. These qualifiers notwithstanding, an examination of the adolescents’ stories reveals that there are aspects of this study that are reflected in the literature on adolescents at risk for Huntington’s Disease and in the theory of identity development of adolescents. Some of the issues that were revealed in the analysis have been addressed in the literature on Huntington’s Disease. Some of the issues have not, and in these areas the findings of this study add significantly to the current knowledge base about adolescents at risk for Huntington’s Disease.

The following analysis focuses on how the results of this study relate to and expand upon what is known so far about adolescents at risk for HD. The two areas of research that are most relevant in this context are the relatively small body of literature on adolescents at risk for HD, and the larger body of literature on adolescent identity development. References to other related literature are included as appropriate.

Adolescents At Risk for Huntington’s Disease

As indicated in Chapter II, there is only a small amount of research that addresses the experience of adolescents at risk for HD. Much of the literature that does exist in this area has not focused specifically on adolescents but has been based on studies of the familial implications of HD generally, and the implications of predictive testing on the family. Moreover, only a few of the published studies that have focused on adolescents at risk for HD have used qualitative methodology (Power, 1984; Korer & Fitzsimmons, 1987). None of the studies have taken a phenomenological approach. Little is known, therefore, about the adolescent’s distinctive or subjective experience of being at risk in a family with Huntington’s Disease.
Naming the Legacy: Understanding and Misunderstanding

Huntington’s Disease became a focus for the adolescents in this study when they were informed that their parent had a disease that would affect their functioning. As with the Power’s study (1977), even when the adolescents were informed that it was a genetic disorder and that they were also at risk, what they heard and what concerned them were the implications for their parent. A majority of the adolescents could only hear that their parent would die. For the two adolescents who had only known their parent as symptomatic, the disease was not an identifying factor of their parent until the disease was named. After this, they began to identify the symptoms and to recognize why their parent was different from other parents.

What caused the adolescents more distress was knowing that there was “something wrong” and not being told what it was. This experience supports the findings of Power (1984), who discovered that when information was not communicated in a family, feelings of anxiety, anger, and insecurity became more pronounced among family members. The message the adolescents in this research received from the adults in the family was that there was a “secret.” They were not allowed to know the secret, they were not allowed to ask about the secret, and they knew the secret affected the family in some way. To ask would violate an implicit family rule that they had learned from observing adult behaviour around secrets.

The adolescents spoke of developing keen observational skills in the process of investigating the mystery around the secrets, and in the end it was those very skills that assisted them in understanding the mystery once the secret was disclosed. These observational skills were helpful in deciphering which of the behaviours of their affected parent were symptoms of Huntington’s Disease and which were not. Moreover, the development of these observational skills was identified as one of the strengths they had received as a result of living in a family with HD. It was a strength that some of them carried with them into later adolescence.
The current analysis highlights the intersection of adolescents at risk for HD and adolescent development. Adolescents construct their own cognitive worlds; they adapt their thinking to include new ideas because additional information furthers understanding (Piaget, 1967). The intuitive sense that underscores children’s insight evolves into Piaget’s preoperational stage (two- to seven years-of-age) in which children begin to represent their sensing of the world with words and images. By seven, children evolve into the concrete operational stage (seven to eleven years of age during which they develop the ability to organize observable and behavioural attributes into cognitive concepts.

The children in this research were informed about a parent having Huntington’s Disease during what Piaget describes as the concrete operational stage of development (seven to eleven years of age). The first part of the experience begins with observing that there is a secret in the family. Procedural knowledge develops (Anderson, 1990), or as Byrnes describes, the “knowing how” (1988a, 1988b). It is during this time that children use their observational skills to begin to identify and conceptualize what they perceive as symptoms of Huntington’s Disease, and to create a category of behaviours that they understand as Huntington’s Disease. As they develop into formal operational thinkers they begin to think more abstractly and are capable of transforming these observational skills and the capacity for understanding their world into more abstract thoughts. It is within this cognitive framework that these skills can be packaged as successful tools in understanding the world. This is the “knowing that” part of conceptual knowledge (Mandler, 1983; Hiebert & LeFevre, 1987; Byrnes, 1988a, 1988b). For example, Marilyn was able to take her observational skills and use them to assist her in adapting to her social world and to solving other problems in her life.
Experiencing the Legacy: Huntington’s Disease in Relation to Relationships

The picture presented in previous studies about the quality of life and relationships in a family with HD is one of impoverishment (Tyler, 1983) and struggles with multiple losses (Kessler & Bloch, 1989; Wexler, 1995; Power 1984), beginning with the loss of the affected parent as the symptoms of HD begin to affect cognitive and physical functioning.

The adolescents in this research described progressive decompensation of their affected parents’ cognitive, emotional, psychological, physical, social, and familial functioning. It was a loss, but not the first loss they recognized. Research on HD families has found that many families deal with HD-related problems like domestic violence, sexual aggressiveness, abuse, and alcoholism (Korer & Fitzsimmons, 1985; Phipps & Desplat, 1884; Yale & Martindale, 1984). The stories of the adolescents in this research demonstrated that problems are not always related to HD; for some of the participants problems were based on issues that predated them learning about HD in the family. Prior to knowledge about HD, the adolescents had experienced parents divorcing, a non-affected parent’s death, two grandfathers’ deaths due to HD, and parental abandonment for an extended period of time. It cannot be determined to what extent these family issues were the result of Huntington’s Disease (including inter-generational effects) or the challenges that face any family. What we do know is that the adolescents described these challenges as issues that were not related to HD or happened prior to knowledge about HD.

There were, however, issues that the adolescents related to Huntington’s Disease. For example, Carla spoke in detail about the loss of her mother in terms of losing a mother she “never knew.” Her mother had been symptomatic all her life. The loss she mourned was for an ideal mother – what she imagined her mother could have been. Kathy, on the other hand, was the eldest of the three-sister sibling group in the research. She had strong memories of her mother as
a healthy and vital person. She discussed how her sisters had never known their mother without symptoms, and she believed that constituted a loss.

The participants did describe an awareness of changes in their affected parent after they were informed of the disease. They began to identify the symptoms of HD and to experience the impact of those symptoms in their lives, particularly with their peers.

What emerged consistently across all the participants was the tendency to judge the affected parent with the same standards with which they judged their non-affected parent, and in the same way that adolescents in general judge their parents. Huntington’s Disease was often not relevant to how the parent with HD was viewed. In fact, in many instances HD was not even considered as a factor in their parent’s behaviour or a criterion by which to judge or understand their parent.

Several factors may have influenced this finding. The adolescents had an historical relationship with their parent throughout childhood that continued to influence their perception of their parent before and after the advent of the symptoms of HD. These findings support the research of Gjerde, Block and Block (1991), who found that the adolescent-parent relationship was affected by the long-term relationship with a parent over the course of childhood. Some of the adolescents in this study were dealing with more immediate issues that impacted their family as well as their relationship with their affected parent. These included the introduction of a new partner for their non-affected parent or the death of a non-affected parent.37

Another factor that affected the adolescents’ view of the parent with HD was the amount of professional and economic support that was available to the family. Those adolescents whose affected parents were primarily cared for by homecare workers or in a homecare facility were free to carry on a relationship with their parent outside the practical constraints imposed by the

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37 There is limited discussion about the non-affected parents’ new partners because of requests from the participants to keep such discussions confidential and removed from the research.
symptoms of HD. Two adolescents in the study were required to be a part of the daily care of their affected parent, yet even those adolescents did not view the symptomatic parent through the “lens” of HD.

As reported previously, the adolescents judge their affected parent using the same criteria that they would use with a non-affected parent. On the one hand they could identify the symptoms of HD and have a concept of the disease; on the other hand it was not the element that defined the identity of the parent. It is a disease that their parent has, but it is not a primary part of how they perceive or judge their parent. For example, though John knew that depression was one of the symptoms of HD, he still judged his father as being capable of resolving his depression and himself as able to delay onset of Huntington’s Disease through preventive measures.

Discussions regarding the non-affected parent emerged in the adolescents’ stories in terms of parent-child interactions in the areas of telling or withholding the secret, bringing another partner into the family, or verifying which symptoms were HD and which symptoms were not HD in their affected parent. They did not describe their non-affected parents’ personality or coping style. For instance, none of the adolescents talked about the non-affected parents’ additional stresses because of the extra demands of HD. It may be the result of the adolescents experiencing their relationship with the non-affected parents as positive and less problematic than the relationship with the affected parents; therefore, they focused more on the problematic behaviour of their affected parent. More importantly, all but one of the non-affected parents were high functioning adults and appeared to make a difference in the quality of life of the adolescents. The adolescents may have perceived the non-affected parents as functioning well and did not observe or acknowledge the impact of the affected parents’ behaviour on the non-affected parents. Only one adolescent spoke of her non-affected parent as providing ongoing assistance in understanding how to cope with the affected parent’s symptoms. This
supports previous studies that found parents of young people at risk for HD felt burdened by the responsibility of giving genetic information to their children. It also demonstrates the possible consequences of keeping family secrets, as well as how these secrets affect family communication.

Sibling relationships in families with Huntington’s Disease have not been widely researched. In this study, the eldest sibling in the two sibling groups reported feeling close to the younger sibling(s), while the younger sibling(s) reported not feeling close and not sharing their feelings about HD with the older sibling. Whether this is the result of family secrets, family communication patterns, or a reflection of typical sibling relationships it illustrates how alone adolescents can feel in an HD family.

All the adolescents voiced some concern about being judged by their peers, and they were selective about who they chose to disclose information to and what information they disclosed. As corroborated in the literature review, friends and friendships are central to developing a healthy sense of self. The adolescents in this research were in an age category for which friends often become more important than family (Deck & Folta, 1989; Sklar & Harley, 1990). As described in Erikson’s (1950) theory of identity development, one of the major tasks of identity formation is experienced within the context of peer relationships (Oltjenbruns, 1996). The uncertainty of how the symptomatic parent might behave creates a sense of vulnerability. The adolescents were embarrassed by the parent’s symptoms, and/or feared the possibility of judgment by peers. The adolescents’ concern was either for their parent with HD or for themselves or both.

All of the adolescents explained Huntington’s Disease using clinical information. When describing HD to their friends, however, they included experiential knowledge of the symptoms that they witnessed in their parent as part of the explanation of HD. The description of their parents’ symptoms was a way to deal with the uncertainty of their parents’ behaviour, and a way
to prepare their friends for any behaviour that might embarrass the adolescent or create the possibility of judgment.

It is important to reiterate that the adolescents did not view their parent only in the context of Huntington’s Disease. Two of the adolescents used HD as an excuse for behaviour that had been embarrassing for them throughout their lives. Having peers regard their parent as angry or inappropriate or “gay” was perceived as far more detrimental than having their parents’ disease status known. The adolescents could handle the possible stigma of Huntington’s Disease better than they could the stigma of having a parent who acted in a way that might be perceived as socially deviant. This finding supports the research in identity development that emphasizes that “acceptance by peers generally, and especially having one or more close friends, may be of crucial importance in a young person’s life” (Coleman and Hendry, 1990:109).

Another way in which the adolescents attempted to protect themselves or their affected parent from the judgment of their peers was to be selective about whom they would bring home. Five of the adolescents did not bring friends home. Comments such as “I try to keep my friends away from my home” and “I try to avoid any contact with other people” illustrate the attempts of the adolescents to control the contact the affected parent would have with their peer group. These efforts at protecting themselves and/or their affected parents from judgment or embarrassment were utilized to assist them in coping with the consequences of having a parent who was symptomatic.

Coping did not always involve keeping friends away from the affected parent. Two of the adolescents wanted to bring their friends home to protect themselves. They believed that having a friend in the home would cause their parent to behave in a less abusive manner than they might if they were alone. The potential for verbal or physical abuse posed a more significant threat than the possible social stigma of the parent’s behaviour with selected friends. This demonstrates a sophisticated approach to problem solving, and supports the theories around critical thinking that
emphasize the adolescent’s ability to not only grasp the deeper meaning of problems but to apply different strategies and perspectives to the problems (Ennis, 1991; Perkins, Jay, & Tishman, 1993; Stanovich, 1993).

Empathy for the parent with Huntington’s Disease was demonstrated by only four of the nine adolescents. Three of those adolescents were from one sibling group, and they all spoke of how they admired their mother’s ability to deal with the challenges of HD. Several factors may have influenced this finding. Although the adolescents’ empathy or lack of empathy could be a reflection of the families coping styles (Power, 1984), other factors seemed to play a role. First, the sisters experienced the most practical support in caring for their mother, with homecare assistance and facilities within the home that allowed for managing the physical symptoms of HD. Secondly, their father was perceived by two of them as being a supportive parent, and they all described their mother as not having mood swings and as being predictable in her behaviour. Thirdly, the introduction of their father’s new partner while the mother was still in the family home confused and upset the girls and may have encouraged them to feel more protective of their mother.

The other adolescent who displayed empathy was Carla. She was able to be sympathetic even though her mother had terrible mood swings and a history of physical and emotional abuse. Carla did not perceive her father as supportive, and felt he violated her right to know about her mother and her mother’s condition by his withholding of information and telling others before telling Carla. Carla did have two experiences in common with the three-sister sibling group. The first was that her mother’s care was managed; her mother was in an extended care home and, consequently, Carla did not have to deal with her mother’s emotional or physical outbursts by herself. The second experience that she had in common was the introduction of her father’s new partner when her parents separated. Her father would not allow her to disclose this information to
her mother. Carla felt very bad for her mother and wanted to protect her from any pain she might feel as a result of her father’s new relationship.

It appears that having professionals manage care of the symptomatic parent was helpful in allowing the opportunity for empathic relationship with that parent. Also, the addition of a new relationship for the non-affected parent, and the secrets involved in the new relationship, were a contributing factor in the adolescents’ ability to have empathy for their affected mothers. For example, Carla knew what it was like to have information withheld about family issues that were significant; she could empathize with her mother’s position of being outside of a circle of knowing that the rest of the family and the social community were a part of.

The other adolescents in this research showed little empathy for their affected parent. This is understandable given that the parents were viewed from the perspective of the adolescents. Adolescents can be viewed as moving through developmental levels within relationships (Paul & White, 1990). The adolescents displayed a self-focus that was more egocentric and concerned with how their parents’ relationship affected themselves and their social functioning in their peer group. Adolescents can understand the facts about HD and identify the symptoms of HD, but to expect empathy from them may be asking for an understanding that is beyond their emotional development and capacity. As cited in Chapter II, the forming of intimate relationships is a developmental task of the early adulthood years (Erikson, 1960). In early adulthood, individuals learn to have self-understanding, as well as consideration of the motivation and needs of others.

**Integrating the Legacy: At the Crossroads of Self and Future Self**

Adolescence is a complex time in human development, and this is supported by the at-risk adolescent experience. Identity development in adolescence represents the first time that physical, cognitive, and social development is advanced to a point where individuals can begin to
sort through childhood identities, as well as their sense of a present and future self, and begin to construct a path toward adulthood.

An examination of self-identity becomes important for two reasons. First, as discussed in the literature review, medical professionals may perceive adolescence as a social problem in and of itself; consequently, normal adolescent behaviour might be regarded as a symptom of disease or pathology (Hill & Fortenberry, 1995). Secondly, many parents are not aware of the normal dimensions of self-understanding and associated behaviours. This lack of knowledge can lead parents to misunderstand the complexity of behavior that is associated with identity development and to possibly construe this behaviour as an early symptom of HD (Korer & Fitzsimmons, 1985; Kessler and Bloch 1989). As demonstrated in Harter’s (1998a) dimension of fluctuating self, the adolescent’s behaviour fluctuates across situations and across time. Parents might perceive these fluctuations as early signs of HD, and misinterpret them as a symptom of the disease rather than as a natural part of development.

Harter’s dimensions also give insight into the ongoing process of development, and demonstrate that it occurs over time and not at one point in adolescence. Although not all of the dimensions of self-understanding help elucidate the impact of Huntington’s Disease in the experience of the adolescents emerging identity, the understanding of this developmental process can provide parents and professionals with important information that can assist in helping the adolescent develop a healthy sense of self.

As viewed through Harter’s first dimension of self-understanding, abstract and idealistic, the adolescents were able to take the concrete experience of their lives as a basis for abstract thought. When judging their parent, whether symptomatic or non-symptomatic, they used their notion of the “ideal parent” as a standard of judgment. The construct of the ideal parent was affected by the adolescents’ experience of the symptomatic parent as well as by the degree of support and communication provided within the family system.
Harter’s second dimension, differentiated self, involves the ability to describe the self with contextual or situational variation. The adolescents in this study demonstrated this dimension in deciding whom they would disclose information to and whom they would withhold information from. Seven of the nine adolescents did not tell their families or medical health professionals about the difficulties they were having. As discussed in the first theme, the adolescents had learned that inherent in having HD in the family was the fact that there were secrets. Those secrets were about HD. They learned not to talk about HD within the family or outside of the family. It became clear that although many of the adolescents appeared to be coping with the ramifications of HD in the family, they were in fact having a difficult time. The withholding of their experience was something that occurred for all the adolescents. The adolescents demonstrated this by acknowledging that the interview for this research was the first time that they had told their story to anyone. The ages of these adolescents ranged from thirteen to nineteen years of age. I find it remarkable to consider what it must be like to hold all the feelings and experiences related to living in a family with HD inside while facing the normal challenges of adolescence. The presence of Huntington’s Disease in the family is not the only challenge facing these adolescents, but from the perspective of self-understanding and identity development the disease places a significant burden on an already difficult period of adolescence.

Adolescent self-understanding also involves social comparison. For the adolescents in this study, social comparison was heightened by the realization that their parent with HD was significantly different than other parents. This difference was demonstrated by how the affected parent behaved, as well as by the parent’s limited participation in the adolescent’s life (e.g. not attending sports events).

The adolescents experienced a more constructive environment for social comparison when they engaged in an adolescent-at-risk support group. Seven of the nine adolescents in this
research attended a support group either through the local HD chapter or at a national HD conference. They found that they could compare themselves to other adolescents who were at risk, and that it normalized their experience of being in a family with HD and at risk for HD. The ability to engage in social comparison in this environment helped free them from the isolation imposed by their silence.

The dimension of self-consciousness implies an introspective part of self-exploration. For the adolescents in this research, it was an aspect of self-discovery that the adolescent would share with friends. They would engage in asking friends about aspects of their self that involved roles and expectations in relationships, but seldom did they speak of their experiences with their affected parent or of their at-risk status. As discussed in Chapter V, only one adolescent spoke of revealing his at-risk status with friends and he found it very difficult. He did not volunteer the information; the disclosure came about as a result of being asked by his friends if he was at risk for Huntington’s Disease.

Carla demonstrated a creative use of self-consciousness and introspection in her own “personal fable” (Lapsley, 1991) about her relationship to a member of the rock group “Spice Girls.” To her they represented a group – a family – with whom she could be connected and who reflected values that she felt represented who she was. Carla is a singer and an entertainer, and believed that her mother could have been as well, were it not for Huntington’s Disease. Carla looked on one particular member of the rock group as a mother-figure. She could share her love of music with her fantasy family where she never could with her mother.

This is an example of the diversity and individuality that adolescents at risk for HD display in constructing creative and positive coping strategies to assist them in developing a healthy sense of self-understanding despite the challenges of HD in their lives. Carla had the ability to create a fantasy world that provided a sense of connection to an imaginary family, and
that helped her with her own sense of loss around a mother she would never know due to the effects of Huntington's Disease.

Within the dimension of real and ideal self, the ability of the adolescent to explore the possibility of a future self emerges (Markus & Nurius, 1986). As described in Chapter VII, the possible self is what individuals might become, what they would like to become, and what they are afraid of becoming. All the adolescents who were fifteen years of age or older spoke of actively thinking about their future possible selves in negative terms; that is, they began to think about a future that included having the gene for HD and being symptomatic. Adolescents in this research under the age of fifteen did not consider negative future possible self or discuss their at-risk status. They were more concerned with the present and with how living in a family with HD affected their social relationships and school activities. They perceived the future as very far away. For some, the future represented a cure for HD that would free them from the negative possible self.

The discussion of this dimension of self-understanding again highlights the intersection of adolescents at risk for HD and adolescent development. The ability to engage in future thoughts about self is a part of normal adolescent development (Erikson, 1968). For adolescents at risk for HD, thoughts about future self often coincide with an affected parent manifesting the symptoms of a disease that graphically represents a potential negative future self.

Two of the adolescents in this study had previously contemplated taking their own lives. (See Carla and Daphne’s stories in Appendix 1.) It was a difficult topic for the adolescents to discuss. One adolescent (Daphne) had actually attempted suicide. She was being treated for depression by a psychiatrist, but at the time of the interview she had decided to stop the medication and try alternative methods to deal with her depression (e.g., meditation). The second adolescent (Carla) revealed for the first time during the interview that she contemplated suicide
and had a plan for how she would kill herself. She was seeing a psychiatrist but acknowledged that she did not reveal to the psychiatrist her thoughts of suicide or her plan.38

The adolescents’ experiences indicate that fifteen years of age and older is a problematic time period; this is reflected in the dimensions of self-protectiveness, the unconscious, and self-integration. During this time adolescents are actively considering their future, and are engaged in activities at school that encourage an exploration of the future. The task of considering future trajectories, part of Erikson’s identity moratorium (1968), placed the future in closer proximity to older adolescents than to adolescents under the age of fifteen. Having a parent who was symptomatic created a future possibility that was not as removed as it was for the younger adolescents. The younger adolescents felt that there would be a cure in the future, or that the future was too far away to worry about. What was important for the younger adolescents was what was happening to their parent and how it affected their relationship with their peers.

Although the older adolescents felt challenged by their future trajectory, a common theme of hope for the future emerged as they described their own future with or without the gene for HD. They talked about their belief that they could fill their lives with rich experiences so that if or when they became symptomatic they would have lived a meaningful life. They revealed that they would live in the present and appreciate what they had rather than worry about the future. They indicated that Huntington’s Disease represented only a part of who they would become, and that the disease would not negate their important self-attributes.

Only three of the adolescents in the study wanted to engage in predictive testing. All were fifteen years of age or older. They felt that predictive testing would give them a sense of control in their lives, and would help alleviate the fear that was engendered by the possibility of a future with Huntington’s Disease. Being from families that maintained secrets and withheld

38 I was able to find a school counsellor that Carla felt comfortable with, and with whom she was willing to talk to about her suicidal ideation. The counsellor had access to additional counselling services if required, and was trained in suicide assessment.
information caused the adolescents to want a sense of control of their own futures. Two adolescents did not want to take the test for fear they could not handle the information if they were to have the gene. As one adolescent said, “The way I see it now is that I’ll think about it when the time comes.”

Two adolescents were uncertain about predictive testing. Marilyn’s first experience with Huntington’s Disease was with her grandfather in the hospital during the end stages of the disease. The image of HD that her grandfather represented terrifies her still, and it is that image that she is confronted with when she considers her future with HD. While being tested for the disease would provide some aspect of control over her life, the information itself represented an undesirable image of the possible future. The thought of testing created a feeling of hopelessness that she did not want to explore. Kathy, on the other hand, felt that she was probably too young to deal with the information, but would consider testing when she was ready to think about having children. She sees HD as a debilitating disease, but one that can be handled with dignity, as demonstrated by her mother’s capacity to manage the disease. Although both girls were uncertain about engaging in predictive testing, they each had experiences with symptomatic family members that became symbolic of how they imagined the disease would manifest for them.

Predictive testing was not expressed as a major consideration for the adolescents at risk. However, the awareness of the implications of the test could be quite profound as evidenced by Marilyn’s fear of having the gene for HD. Kathy, on the other hand, represents a more pragmatic approach to predictive testing; she sees it as providing information that will inform her choice about reproduction. Each was influenced in their decisions by an image of HD that was represented in their affected family members. The remaining four adolescents were either indifferent about testing (believing that there would be a cure) or were in the early stages of adolescence and were focused on more immediate issues. The younger adolescents did not
include predictive testing or the implications of having the gene for HD in their picture of possible future self until asked about predictive testing. Asking the question did not seem to influence their thoughts but served rather to elicit them.

The factors that influenced thinking about predictive testing were the age of the adolescents, adults asking the adolescents about predictive testing, and the need to have control in their lives by choosing whether to access information through predictive testing. Predictive testing represented an action that gave them – often for the first time – control of information about Huntington’s Disease that would allow them to make informed decisions about their future.

Although it varies among individuals, later adolescence is a time when the adolescents begin to consider their future and the possibility of predictive testing. This research indicates that it is during later adolescence that at-risk individuals start to consider their future and the possibility of taking predictive testing. It is also a time when a potential negative future is actualized in their affected parents’ symptoms of HD.

The adolescents’ view of the future illuminated a pattern that was consistent across all of the adolescents’ experience. They all had existential sentiments about the meaning of life and relationships that would not be developmentally expected for their age range. It was a maturity that emerged from not being able to take for granted fundamental aspects of adolescent life, such as relying on parents who were physically healthy and capable of managing the activities of daily living and of providing physical and emotional support for their families. Being in a family with HD placed additional stress on these adolescents, but it also seemed to result in a resilience and maturity beyond their years. As Margaret said,

There are going to be times you’re going to be sad, but you have to deal with it. You can’t let it run your life. Sometimes I felt sorry for myself, but you can’t let it interfere. It may sound like a cliché, but it is just life and you need to live it.
Coping Strategies

As discussed in the literature review, Power (1984) utilized Lazarus’ (1984) coping model to delineate the coping mechanisms of the adolescents in his research into two categories: “problem-focused” and “emotion-focused.” Lazarus argued that well-functioning people use a mixture of problem-focused and emotion-focused coping methods (Lazarus, 1984). The adolescents in this research used both forms of coping mechanisms; they did not display “giving up” behaviour in the form of withdrawal, nor the “unwarranted dependence” that Power found in his research. In this study, the adolescents’ stress could be seen as the threats and challenges that environment placed on them, or it could be viewed as the adolescents’ responses to such threats and challenges. Because debate continues in the literature as to whether stress is the threatening event in the adolescent’s world or a response to those events, a broad definition of stress is used in this research: Stress is the response of individuals to the circumstances and events – stressors – that threaten them and tax their coping abilities.

Many factors big and small can produce stress in the lives of adolescents. Clearly, being at risk for HD and living in a family with HD contributed significantly to the stress load of the adolescents in this research. Yet despite the challenges represented by HD, what emerged from the adolescents’ stories was a theme of resilience. Resilience was reflected in the creative coping strategies of the adolescents. Just as stress can be a difficult term to define, resilience can be even more difficult. As Vaillant (1993:3) states, “We all know perfectly well what resilience means until we listen to someone else try to define it.” Resilience has varied meanings across existing research studies; for the purpose of this research, resilience is defined as positive changes in maintaining active or latent coping and adaptation capacities through various mechanisms that may not be immediately apparent but become evident over time (Foster, 1997). The latter point is important because it stresses the necessity of carefully considering the time at which we
measure resilience, and of the necessity of remembering that one’s resilience may change over
time and in different domains.

Resilience can be described in three developmental phases: child, adolescent, and adult. In children, resiliencies appear as uniform, non-goal oriented, intuitively motivated behaviour. In adolescents, these behaviours sharpen and become deliberate. In adults, they broaden and deepen, becoming an enduring part of the self. For instance, as stated in Chapter V, insight begins with sensing in childhood, becomes knowing in adolescence, and matures into understanding in adulthood.

The adolescents in this research demonstrated a variety of ways of coping. These coping techniques involved three different types of behaviour: personal, interpersonal, and group. All of the coping mechanisms were reported as helpful, although certain behaviour – anger, smoking, withholding information, for example – would not necessarily appear as positive.

Personal behaviour

The first category of coping concerned activities or ways of thinking that involved the adolescent only. Two of the coping skills – observing and avoiding – involved skills that the adolescents reported developing over time. Three adolescents spoke in detailed terms about how observing family members and peers helped them learn how to cope with their lives generally and with ongoing problems related to their symptomatic parent specifically. For instance, Marilyn talks of observing others all her life because she was “never told anything.” Since childhood she has consciously watched family members in order to figure out the secrets she sensed within her family. She reported using the same skills to watch peers whom she felt coped well. In the first instance, she observed to gain knowledge; in the second instance she observed in order to learn and then emulate the coping skills used by her peers. She spoke about her observation skills as one of the assets she developed as a result of living in a family with HD.
Four of the adolescents referred to avoidance as a way to protect themselves from the inevitable conflict or erratic behaviour of a symptomatic parent. They felt powerless to effect any change, and felt that if they could leave the situation it would either resolve itself in time or a person with more authority or power would be able to change the affected parents’ behaviour. These coping mechanisms are conscious efforts on the part of the adolescents to control or relieve on-going pressures in the family. The adolescents who reported these coping techniques were the same adolescents who spoke of living in a family where there were secrets and where they learned not to ask questions about the tensions they perceived in the family.

The adolescents in this study also employed anger as a coping mechanism. Anger is an important feeling. It gives us a signal that something is not right. Adolescents get angry with their parents for many of the same reasons they get angry with their peers. They are frustrated because they cannot do what they want, or get something they want. They also get angry in response to parents’ demands, or in response to parents’ anger. The adolescents in this study got angry with their parents for particular reasons. They felt afraid of certain events or of whatever they suspected was going on, and they used the anger to express or cover up those concerns. Rather than feel helpless, adolescents will often become angry. For instance, Carla was angry at her father for not informing her of what was happening with her mother who was symptomatic. Her mother was taken to the hospital after a frightening event in the home, and Carla did not know when she would see her again. Her father did not inform her of what was happening, or if her mother would return home. She felt she was always the last to know what was going on in the family, and she was never sure if she had all the facts about her mother’s well-being. She was constantly using angry terms to talk about her father and the fact that he would not share his frustrations with her. She kept her feelings inside and appeared depressed. When exploring her feelings, she acknowledged that she felt depressed and at times suicidal.
Anger can also be a way that adolescents attempt to gain some control and a sense of power over their lives. As stated in Chapter VI, Marilyn used anger toward her affected father as a way to frustrate him. His expression of frustration served to reveal to Marilyn that his disease kept him from dealing with life normally.

All but one adolescent in this study seemed to be angry. The anger was most often directed toward the non-affected parent. As mentioned, the non-affected parents in this research seemed to be responsible for all the activities of daily living, so it was understandable that the adolescents assumed that the healthy parent was responsible for any difficulties that arose in the family due to the additional burdens placed on the family by HD.

Another factor in how the adolescents managed their anger was how the parent with HD handled their own anger. Six of the adolescents were afraid of their affected parent's anger, and were afraid to show their emotions or get angry for fear the parent would harm them. Two adolescents spoke of physical abuse by their affected fathers. Both Emma and Stephanie spoke of abuse that occurred in the past. Stephanie's father began to take new medication after losing his temper for the second time and striking her with his cane. Stephanie commented that while the medication completely changed her father's temperament she still could "not trust him." Emma reported that while her father was no longer physically abusive he continued to be verbally abusive. Carla reported being afraid of her mother's outbursts, but it was only when her mother's symptoms progressed that she began to be afraid for her own safety with mother. She recalled in detail one evening when she stayed with her mother and her mother chased her with a bowl that she threatened to "push down my throat."

The burden of appropriate responses to emotions or emotional situations falls on the non-symptomatic parent. Acknowledging feelings of anger or sadness is important; the adolescents in this research needed to be taught how to have feelings, that feelings are normal in stressful situations, and that you don't take feelings out on your family. They also needed assistance in
learning how to protect themselves when a symptomatic parent acted inappropriately, in a threatening manner, or in a physically abusive manner.

The activities that assisted the adolescents when they felt overwhelmed ranged from smoking cigarettes to writing in a journal. Music is a powerful vehicle for the expression of feeling, and, as with most adolescents, music played an important part in all of the at-risk adolescents’ lives. All the adolescents found that listening to music gave them pleasure and helped them to relax and reflect on their feelings. Some of the adolescents played instruments, and they reported that this provided distraction and peace of mind. Two adolescents wrote songs about their feelings and about HD. Carla shared a song she wrote called “If I promise not to feel this pain, will I see you again?” It was a poignant song about the ravages of HD that stole her mother “away.”

For John, the process of predictive testing became a coping mechanism in that it provided a way for him to deal with the stress of uncertainty. As discussed in the previous chapter, John decided to engage in testing because he felt that he was a “pessimist” and would always think that he had HD. Taking the test would give him a definitive answer and allow him to move in specific directions in his life regardless of the test result. John also believed that he would access information that would lead to some medical development that might delay the onset of symptoms by “ten years.” Regardless of whether there is any reason to believe that the onset of symptoms could be delayed, the idea of predictive testing and the possibility of some means of delaying onset of symptoms helped John cope with his at-risk status.

Interpersonal behaviour

The second type of coping behaviour involves interpersonal relationships. All of the adolescents cited talking to a best friend(s) and/or a boyfriend or girlfriend. Not all of the adolescents believed that their friends could “really understand” but rather believed that they
could provide support and understanding when they could not find it in their family. What stood out for me was that only three of the adolescents felt that they could share their deeper feelings with their family. Kathy and Daphne felt they could speak to their siblings. Marilyn felt she could speak with her mother. In other cases, however, the extra demands on the non-symptomatic parent could result in there being little time to attend to the emotional needs of the family. Additionally, most of the adolescents in this research spoke about family secrets and exclusion from family communication. The lack of communication among siblings may be a reflection of the family's style of interaction, or it may be owing to differences in the siblings' age, gender, or developmental stages. Although Kathy and Daphne felt that they had good communication with their siblings, the younger siblings in these two sibling groups reported a different view of the communication; in fact, they felt that there was little or no communication between the siblings.

An important source of support came from mentors. There are many interpretations of what would constitute a mentor. For the adolescents, a mentor was a person who really cared, was there to help, and treated them with respect. The adolescents perceived their mentors as having competence and knowledge that they did not have, and it was important to them that their mentors were able to share that knowledge with them. Five adolescents sought out mentors to assist them in dealing with the everyday experiences of life as well as with family issues that arose from HD. The mentors included a boss, a grandmother, the mother of a best friend, a social worker, and a teacher. None of the adolescents felt comfortable talking to doctors, psychologists, psychiatrists, or genetic counsellors.

**Group behaviour**

The third category of coping involved participation in support group activities. Eight of the adolescents were involved or had been involved in some group aspect of the Huntington
Society. The activities included attending a national HD conference, an adult-at-risk-for-HD support group at the national conference, a monthly meeting of a local HD chapter, and an adolescent-at-risk-for-HD support group. Only one adolescent did not become involved in some way with the HD Society. The level of involvement of the adolescents with the HD Society may be a reflection of how I recruited adolescents for this research, as I met all of the adolescents through my activities with the Society or with the HD Predictive Testing Clinic, or I was introduced to the adolescents through the Society’s social worker. Of the eight adolescents that attended HD Society functions, only one adolescent found that it was a hindrance to coping. In her case she found it was too difficult to be with people who reflected her own sense of “mortality.”

Summary

Chapter VIII presented the overarching structure of the adolescent experience of living in a family with HD and at risk for HD. Three themes were identified, and a common story of the co-participants’ experiences was presented.

The first theme, Naming the Legacy, described the process of the adolescent’s learning about HD and being at risk for HD. Before the adolescents were told about the disease they had intuited that there was something “different” or “wrong” in the family. The adolescents were more distressed about the “secret” than the disclosure of the disease. Having a secret in the family created an understanding that there was something unspoken that they could not talk or ask about and that affected the family. This situation created the opportunity for the adolescents to develop observational skills as a means of trying to discern what the secret might be. They continued to use these skills after the secret was disclosed to help them understand what were the symptoms of HD and also as a tool for understanding situations outside of the family.
The second theme, Experiencing the Legacy, described the adolescents' experience of living with a symptomatic parent and the impact this had familial and peer relationships. The problems that the adolescents experienced with the affected parent were not always related to HD but rather to issues that predated knowledge of the illness and that were not dissimilar to what many families face. The adolescents described an awareness of the changes in their affected parent after they were told about the disease, and began to experience the impact of those symptoms in their lives and in their peer relationships. Most of the adolescents did not communicate with their siblings about their concerns or the challenges of living in the family with HD, but they all had a friend(s) that they shared their thoughts with. The adolescents were embarrassed by their parent's symptoms, and/or feared the possibility of judgment by friends. The adolescents explained HD and the symptoms associated with it to their friends, but many did not bring friends home in order to protect their parent or themselves from judgement. Empathy for the affected parent was shown by only four of the adolescents. Managed care of the symptomatic parent was one of the variables that allowed the opportunity for an empathic relationship with the affected parent.

The third theme, Integrating the Legacy, explored the consequence of living with HD and at risk for HD on adolescent self-understanding and identity development. Harter's (1990a, 1990b) dimensions of self-understanding helped elucidate the impact of HD in the experience of the adolescent's emerging identity. It is within the dimension of real and ideal self that the ability of the adolescents to explore the possibility of a future self emerged. The adolescents fifteen years of age or older began to actively think about their future possible selves as holding the possibility of having the gene for HD, and three of the adolescents in this age range wanted to engage in predictive testing.

Being in a family with HD seemed to result in a resilience and maturity beyond their years, even though HD placed an additional stress in their lives. The adolescents in this research
reflected resilience in the variety of ways they learned to cope with living at risk in a family with HD. These coping techniques involved personal, interpersonal, and group behaviour. All of the coping mechanisms were reported as helpful. The first category of coping concerned activities or ways of thinking that involved the adolescent only. Two of the coping skills, observing and avoiding, involved skills that the adolescents reported developing over time. The second type of coping behaviour was one that involved interpersonal relationships. All of the adolescents cited talking to a best friend(s) and/or a boyfriend. An important source of support came from mentors. The third category of coping concerned the adolescent participating in support group activities. Eight of the adolescents were involved or had been involved in some aspect of the Huntington Society.
Stories are not replicable because our lives are unique. Our uniqueness is what gives us value and meaning. Yet in the telling of stories we also learn what makes us similar, what connects us all, what helps us transcend the isolation that separates us from each other and from ourselves (Remen, 1996:xv).

This research has been concerned with illuminating the experience of adolescents at risk for Huntington’s Disease, and has been aimed at understanding the meaning of the experience. Phenomenology guided this investigation and provided a structure for revealing the phenomena to the researcher. As Van Manen (1990) has noted, one cannot reflect on experience while living through it; phenomenological reflection is, therefore, retrospective in nature. The adolescents’ stories, my research, my experiences in the HD community, and my work in the clinical setting of predictive testing for HD have all provided the background through which the phenomena of the adolescent experience of being at risk for HD was viewed and distilled. The aim for understanding – not only for the researcher but also for parents of at-risk adolescents and professionals who work with HD families – is reflected in this chapter. The chapter is divided into three sections: (1) Theoretical Contributions (2) Future Research, and (3) Implications for Practice.

Theoretical Contributions

This dissertation challenges the view that adolescence is a time of rebellion, crisis, pathology, and deviance (Hall, 1904; Hill & Fortenberry, 1992; Fabrega Jr. & Miller, 1995). This study has developed a vision of adolescence as a time of evaluation, decision-making, commitment, and of carving out a place in the world. Development is not entirely external and other-determined and, likewise, not entirely internal and self-generated. Adolescents bring
certain developmental capacities to any situation and act in the situation. At the same time, however, they interact with others who offer their own versions of the world, which adolescents learn from and sometimes adopt for their own. Adolescents are not a homogeneous group. Different portrayals of adolescence emerge, depending on the particular group of adolescents being described. The group of adolescents that this research describes are adolescents at-risk for HD and living in a family with HD. Each adolescent is unique, yet their shared experience of living with and at risk for HD contribute to our current knowledge base about adolescence in general and at-risk adolescents in particular.

Adolescence is a time when individuals engage in the process of identity formation. It is also a time when cognitive abilities develop and assist adolescents in understanding their world. Piaget’s theory of cognitive development was reflected in this research (1961). Adolescence is the time that Piaget calls “the formal operational stage.” Although it is important to recognize adolescents’ capacity to engage in more abstract thought at this stage, the adolescents in this research were recognizing differences in the family at a younger stage of development. The intuitive sense that underscores children’s insight is indicative of Piaget’s “preoperational stage” (age seven to eleven). It was during this time that the participants in this study described intuiting that something was “wrong” or “different” in the family, and this recognition set up the beginning of a process of learning about Huntington’s Disease. When as adolescents they were told that their parent had HD and that they were at risk for the disease, they were, in essence, offered the “piece” that completed the puzzle created by their intuition.

The developmental nature of cognitive abilities and its impact on understanding of self and others influenced the adolescents’ relationships with their parents and the way that they perceived and judged their parent. The adolescent-parent relationship was affected by the relationship developed with the parent over the course of childhood. Although the adolescents described progressive decompensation of their affected parents’ cognitive, emotional,
psychological, physical, social, and familial functioning, it occurred within the on-going process
of individual and relationship development. Huntington’s Disease was a part of who the parent
was becoming but was not overly significant to how adolescents viewed or understood their
parent.

Parent’s symptomatic behaviour became a concern when the adolescents were engaged in
developing self-understanding and peer relationships. This occurred during a time when
adolescents were involved in what Erikson refers to as identity formation (1968). Consequently,
as the symptoms of the parents progressively decompensated, the adolescents were progressively
developing cognitive, emotional, psychological, physical, social, and familial functioning.

The non-affected parent was primarily discussed as the keeper of the family secret or the
teller of the family secret. The adolescents did not communicate with their non-affected parent
about their affected parent or about their own concerns about being at risk for HD. Power’s
(1977) suggestion that worry for the affected parents resulted in over-dependence on the non-
affected parents was not evidenced in this detailed study of nine adolescents.

In this study, sibling relationships and birth order were not consistent with general
theories on sibling relationships, although it must be noted that sibling relationships in HD
families are poorly described in published research. The general theory proposes that adolescent
sibling relationships are emotionally charged, marked by conflict and rivalry, but also provide
nurturance and social support (Lempers & Clark-Lempers, 1992). Adolescent siblings can also
act as communication partners (Vandell, 1987). The two sibling groups in this research did not
engage in these activities, although the oldest siblings in each of the groups reported good
communication with the younger siblings. The siblings operated independently of each other and
did not turn to each other for help, teaching, playing, or emotional support.

Birth order did impact the adolescent’s relationship to the affected parent. It is unclear
how this relates to parent-child relationships in general due to limited research in this area
(Suitor & Pillemer, 2000). However, the older the sibling, the stronger the likelihood of experiencing a parent free of the symptoms of HD. The only parent that the younger siblings knew was a parent with the symptoms of HD. Consequently, they did not perceive a past loss of not knowing the mother that their elder sibling knew; rather, they experienced loss in terms of the progressive course of HD in their present lives.

The adolescents in this research stressed the importance of friends and friendships. As demonstrated in the literature review, friends can become more important than family (Deck & Folta, 1989; Sklar & Harley, 1990). Given that a major task of identity formation is experienced within the context of peer relationships (Oltjenbruns, 1996) and that communication with parents and siblings was limited in the families of these adolescents, it was not surprising that friends were perceived as the primary emotional support for these adolescents. All the adolescents were concerned about being judged by their peers, and were selective about who they chose to disclose information to and what information they disclosed. What was illuminating was how the adolescents used HD as an excuse for an affected parent’s behaviour (e.g. anger, effeminate mannerisms) that was not related to Huntington’s Disease. The affected parent’s symptoms were a source of potential stress with peers, but it was not the only behaviour that stressed the adolescents.

Although the adolescents could understand the facts about HD and identify the symptoms of HD, only four of the adolescents displayed empathy for their parent with HD. As in Power’s study (1977), the adolescents were concerned about their parents’ possible death more than their own risk status. Yet this worry did not translate into empathy in the daily experience of living. This may be explained by the adolescents developmental stage, as it is not until early adulthood years that individuals learn to have consideration of the motivation and needs of others in intimate relationships (Erikson, 1968). It could also be that empathy is intermittent when an
adolescent experiences the daily disruptions of the affected parent's behaviour and the inability of the parent to meet the needs of the adolescent.

The ability of the adolescent to explore the possibility of a future self is a normal part of the development of self-understanding (Harter, 1998a, 1998b; Markus & Nurius, 1986). The presence of Huntington's Disease in the family created an additional stress on this aspect of identity development and self-understanding for the at-risk adolescents. The thoughts of future self coincided with an affected parent manifesting the symptoms of a disease that graphically represented a potential negative future self. Two adolescents in the research had suicidal ideation, with one adolescent unsuccessfully attempting suicide. Each reflected on their fear of a future that would be similar to their affected parent. Kessler (1993) reported that suicide in the HD family system was far more prevalent than in the normal population. Due to the methodology used in this research, the discovery of the suicidal ideation of two of these nine adolescents and the relationship of these actions to their fear of HD in the future cannot be generalized.

The adolescents did not actively engage in considering possible future selves until they were fifteen years of age or older. The younger adolescents were far more concerned with the impact of HD on their lives in the present. Even when I asked them about the future, they did not speak about being at risk or a future that included having the gene for HD. They did speak about believing that a cure for HD would be found by the time they reached the age of onset. This optimism can be a reflection of their cognitive development or an act of avoiding confronting the risk for the present.

Whether the actions of these adolescents are viewed as developmental or coping mechanisms, and despite the fact that two adolescents had contemplated or attempted suicide, the adolescents attempted to hold on to a theme of hope whether they described their future with or without the gene for HD. Research demonstrates that three factors often appear to help children
and adolescents become resilient to stress: (1) cognitive skills and positive responsiveness to others; (2) families marked by warmth, cohesion, and a caring adult(s); and (3) the presence of some source of external support (Garmezy 1985; 1993; Werner, 1989). Most of the adolescents in this research did not have all three of these factors influencing their lives.

Only one adolescent was adamant about engaging in predictive testing. This finding may not be surprising, since only 5 to 15% of the at-risk adult population engages in predictive testing (Richards, 1996; World Federation of Neurology Research Group on Huntington’s Disease, 1993), although how adult behaviour compares to adolescent reports is difficult to assess. Gender may have played a role in the decision, as this adolescent was the lone male in the study. However, the fact that he was the only male taking part in the research meant that the impact of gender could not be fully explored. The factors that did influence thinking about predictive testing were age, adults asking the adolescents about predictive testing, and the adolescents’ need for perceived control in their lives through accessing information. What was reflected in the adolescents’ experience was that the interest in predictive testing was not only a result of developmental age but also a consequence of adults withholding or presenting information about predictive testing.

Through an exploration of self-identity and future self this research illuminated how the experience of the adolescent at risk for HD is best characterized as an individual process that is influenced by the cognitive, developmental, and socio-cultural context of the adolescent’s life. An unexpected pattern did emerge from the adolescents’ experiences: they all displayed a maturity that was displayed through their existential thoughts about the meaning of life and relationships and that would not be developmentally expected for their age range. Being in a family with HD placed additional stress on these adolescents but it also resulted in a resilience and maturity beyond their years. This discovery points to the need for a collaboration between developmental psychologists, pediatricians, genetic counsellors, anthropologists, psychiatrists,
and other scholars and professionals in order to further illuminate the complex state of being an adolescent at risk for HD at the cross roads of self identity and future self. 39

**Future Research**

Adolescents in this research intuited that something was “different” or “wrong” in the family as early as five years of age. They described the process as intuiting that something was happening but not being able to corroborate the intuition through cognitive reasoning. The role of intuition in cognitive development, and its impact on understanding as a child develops, needs further exploration. Of particular interest is how intuition impacts adolescents at risk for HD and other genetic disorders. Psycho-social research into child and adolescent understanding of genetic disease and at-risk status that looks at the individual’s experience of intuition, the context in which the intuition occurs, and the impact of the intuition on later stages of learning about HD would help elucidate this complicated process.

The adolescents in this research experienced more distress from their unsubstantiated sense that something was “wrong” than from receiving information about Huntington’s Disease. Qualitative research that explores the experience of “secrets” in the family and the impact of these secrets on inter-family relationships, family communication, and adolescent communication both within and outside of the family would contribute to a better understanding of the role of cognitive understanding and information on mitigating stress based on experiences that are otherwise intuitively grasped and not explicitly informed. The proposed research would assist families and professionals in developing effective ways to communicate disturbing information about genetic disease to children and adolescents as well as alleviate undue stress in a family already challenged by the impact of HD. An exploration of the effects of third-party

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39 This finding has implications for counselling and decisions about predictive testing and will be discussed in the last section on Implications for Practice.
Interveners on family dynamics and individual coping in a family with HD would provide salient professional intervention strategies. Interventions that could be explored include use of mentors, family therapy, and psycho-educational groups.

The adolescents in this research demonstrated a shift in self-understanding regarding their possible future selves at fifteen years of age and older. The shift in awareness represents an important aspect of cognitive development (Erikson, 1968). The task of integrating varying self-conceptions is coupled with the emergence of formal operational thought that presses for integration and the development of a consistent, coherent theory of self (Harter, 1990b). This capacity for abstract thought and its impact on self-understanding can be significant, especially when an adolescent has the additional stress of incorporating being at risk for a genetic disease as a possible future trajectory. A comparative study of younger and older adolescents across genetic diseases would provide helpful strategies on how to disclose information about genetic risk to adolescents at different times of development. Also pertinent is the impact of this information on adolescent decision-making across the various cognitive stages of adolescent development, and a deeper understanding of how adolescents formulate the idea of risk and the implication of that understanding on their notions of possible future self.

There was little discussion of extended family members in all of the interviews with the adolescents. For some of the adolescents, relationships with their grandparents were powerful influences on how they perceived HD, how they coped with their affected parent, and how they viewed their own risk status. A future exploration of the intergenerational effects of Huntington’s Disease would help illuminate the lived experience of being in a family with a genetic disease and its impact over generations. Are communication patterns, secrets, folklore, and misunderstandings a part of the legacy of HD that is passed on through generations? Can grandparents and aunts and uncles become mentors for adolescents as they begin to learn about
HD, or are the intergenerational effects of HD too challenging and therefore contribute to a lack of support among extended family members?

As previously noted, there has been little research into sibling relationships in families with Huntington's Disease. This study found that there was a discrepancy among siblings about perceived communication patterns. The older siblings believed that communication was open between the siblings, whereas younger siblings reported closed communication among their siblings. What accounts for this lack of inter-subjectivity? Are younger siblings unable to recognize older sibling support? Is this discrepancy a reflection of family communication styles and the keeping of "secrets"? How can sibling communication be supported? A study with a larger population of sibling groups that examines adolescent communication styles within sibling groups, the effect of birth order and gender on communication, and the influence of parental communication style on sibling communication would illuminate our understanding of the nature of sibling relationships in the context of hereditary illness.

Given that the adolescents in this research did not communicate their feelings or concerns to their parents or siblings, friends played an important role in supporting the at-risk adolescents. A particularly salient issue for further exploration would be the nature of friendship relationships as protective functions. In what ways do the relationships with friends assist the at-risk adolescents in developing relationship competencies such as perspective, empathy, and social problem solving? How do at-risk adolescents decide whom they will talk to about the family "secret"? In this research, the adolescents did not discuss their risk status with their friends. Could this have been a result of denial of their risk status, or was it a deliberate decision? If so, how are these decisions made to disclose or not to disclose to friends?

The resilience and maturity that the adolescents in this research demonstrated suggests that resiliency would be a useful topic to explore with adolescents at risk for HD. As discussed in the section on implications for theory, research has demonstrated that three factors often appear
to help children and adolescents become resilient to stress: (1) cognitive skills and positive responsiveness to others; (2) families marked by warmth, cohesions, and a caring adult(s); and (3) the presence of some source of external support (Garmezy 1985; 1993; Werner, 1989). Most of the adolescents in this research did not experience all three of these factors. Further research should explore how consistent this finding is for adolescents at risk for HD and for adolescents with other genetic disorders. What is the origin of this resilience in adolescents at risk for genetic diseases and does it persist over time? What are the coping mechanisms employed by these adolescents, and how effective are they in supporting their resilience and capacity to cope with the challenges of living in a family with genetic disease?

**Implications for Practice**

**Assisting Adolescent Coping**

It is important that both parents and professionals recognize adolescent coping mechanisms as conscious efforts to control or relieve on-going pressures in the family. Parents and professionals would enhance their effectiveness if they were actively curious about the adolescents' coping techniques, and if they identified and validated the positive techniques they witnessed.

The activities that assisted the adolescents when they felt overwhelmed included smoking cigarettes, writing in a journal, and listening to or creating music. Parents and therapists could, for example, ask adolescents to share a few of their favourite songs, as the songs and lyrics often provide remarkable insight into the kinds of emotions and values they have. A discussion about the music becomes a means of expression of feelings about self and others. An example of this is the song that Carla shared with me about her mother's advanced HD (discussed in Chapter VIII). I asked her about her favourite music and if she would share it with me. What she shared was her own song that reflected the helplessness and despair of an adolescent seeing her mother in the
latter stages of HD. We were able to discuss these feelings, and to explore how she could get support from a counsellor at school to learn how to manage her feelings. Had I been the professional working with her, it would have been an opportunity for us to begin to identify and label her feelings, which would help in normalizing her reactions and assist her in handling her profound loss.

Two further ways to support adolescent coping would be to encourage healthcare providers to let adolescents know that they are there for them and are approachable, and also to inform parents about the vital role they can play as mentors to their adolescent children. Beier et al. (2000) found that there is a relationship between adolescents who have mentors and less participation in certain risk behaviours. They also assessed the impact of non-parental mentors and parental mentors versus no mentors and found that adolescents with either type of mentor were likely to be at less risk for high-risk behaviours than adolescents who did not have a mentor. Given that in many families with Huntington’s Disease the non-symptomatic parent may be overburdened with the responsibilities of the family, Beier’s findings suggest that finding non-parental mentors for at-risk adolescents would assist the adolescents and their families.

A final recommendation for supporting adolescent coping is to encourage the use of peer support groups. Eight of the adolescents who were involved or had been involved in support groups with the Huntington Society found the experience to be helpful and a source of validation and normalization of their experience of being at risk and in a family with HD. Peer support activities might include attending support groups at national HD conferences, monthly meetings of their local HD chapter, or adolescent-at-risk-for-HD support groups.

Assisting Adolescents with a Symptomatic Parent

All of the adolescents in this research reported challenges in dealing with their parent who had HD. Four concerns that were raised by all the adolescents are addressed here. Parents
who are involved in helping an adolescent deal with a symptomatic parent may find these concerns enlightening.

1) Children are not responsible for the affected parent’s behaviour.

All of the adolescents found it difficult to understand their affected parents’ behaviour as well as the responses they received from their affected parent. As described in the research, the adolescents learned about HD through both factual and experiential knowledge. They, like the rest of the family, needed time and experience to understand what were symptoms of HD and what were not symptoms of HD. The adolescents’ confusion about what were symptoms and what to expect from affected parents could lead them to assume that they were responsible for their affected parent’s distress or anger. It is important that parents assure their children that they are not responsible for the behaviour of the parent. It is also important that the parents assist them in understanding the facts about HD, including the behaviours that are associated with it.

2) Huntington’s Disease can represent a stigma in public situations or with friends.

As Huntington’s Disease progresses, the symptoms of an affected parent become more pronounced. The choreic movements of the affected parents in this study were the cause of a variety of public responses that created concern for the adolescents. As discussed in Chapter VI, the adolescents experienced the reactions of friends and the public as a source of embarrassment and judgment for both themselves and their parents. They engaged in complicated social strategies designed to alleviate the potential stigma of their affected parent’s behaviour. These strategies included not allowing friends to visit their house, or avoiding public situations with their parents. For some of the adolescents, the response from public situations became less of a concern the more they understood and accepted their parent’s behaviour as an inevitable part of the progression of HD. It is important for parents and professionals to acknowledge that a public reaction to affected parents might exist and to discuss how people are often afraid of behaviour
that they do not understand. Non-affected parents can offer some suggestions based on how they handle situations that arise when there is a social reaction to an affected individual’s behaviour.

3) It is difficult to handle an affected parent’s mood swings and/or behaviour.

Although mood swings are a probable symptom of HD, not all people who are symptomatic experience mood swings. Three of the adolescents in this study who were sisters did not experience their affected mother as having mood swings. Six of the adolescents had problems understanding how to handle their affected parent’s mood swings, and felt there was little they could do that would be perceived as helpful to the parent with HD. Many of the adolescents were not willing to share their frustrations and concerns about their parent with HD because they did not want to place an additional burden on the non-affected parent. It is important for parents to check in with their children about how they may be handling the personality changes associated with HD. They may need to pay attention to any physical or emotional signs of anxiety that the adolescent displays, such as stomach aches, headaches, problems sleeping, or problems concentrating.

Because of the progressive nature of HD, it can be difficult to know if and when individuals with HD are physically and mentally fit to take on the role of sole parent. When communication breaks down between the adolescent and the affected parent, it may well be time to consider alternatives to taking on the task of parenting the children in the family by themselves. I recommend getting assistance with parenting from relatives, support workers, or live-in help. The consequences of erratic behaviour and angry outbursts can be damaging not only to the adolescent but to other family members as well. Emma and Daphne demonstrate in their stories how difficult and potentially dangerous it was for them to assume a helping role for the parent with HD. Daphne was afraid of her father’s anger yet felt it was disloyal for her to discuss it with her mother. She was torn between protecting herself from her father’s behaviour and protecting him from being seen as a “bad” parent.
4) It is normal to experience a range of feelings and concerns regarding risk status.

Although the adolescents in the research shared a common theme of hope for their future they also expressed feelings that created stress and worry. As stated in the literature review, researchers are reluctant to talk to adolescents about their risk status or their thoughts about the future for fear of bringing up distressing and problematic feelings for the adolescents. All of the adolescents told their story of living in a family with HD for the first time in this research and found it helpful to discuss their risk status with me. None of the adolescents felt that they could discuss these topics with their parents or siblings. As stated earlier in this chapter, many of the adolescents did not share their concerns or anxieties with their non-affected parent for fear of adding to the already overwhelming burden of managing the family. The research highlighted the need for adolescents to have the opportunity to discuss their fears and concerns about their risk status and their future with their family or trained counsellors or therapists.

Implications for Genetic Counselling

The testing of individuals before the age of majority for hereditary late-onset diseases has been a topic of much debate. Guidelines and directives published by several medical professional organizations consider genetic testing of adolescents to be unethical because adolescents are presumed unable to benefit from the information in a manner that would justify the attendant risks. In 1994, the International Huntington Association and the World Federation of Neurology Research Group on Huntington’s Chorea published its guidelines concerning genetic testing for HD (Clinical Genetics Society, 1994). They explicitly recommended that minors not be tested.

A working party was convened by the Clinical Genetics Society to engage in global discussions on the question of genetic testing of minors (1994). The working party recommended not to test any healthy persons before adulthood for diseases that do not fulfill
one or both of the following criteria: (1) manifestation in childhood, and (2) the presence of preventive or therapeutic measures initiated in childhood (1994).

The American Society of Human Genetics and the American College of Medical Genetics (ASHG/ACMG) published a joint report on genetic testing in children and adolescents in which they state that “if medical or psychological benefits of a genetic test will not accrue until adulthood, as in the case of ... adult-onset diseases, genetic testing generally should be deferred” (1995:1240). They admit, “exceptions to this principle might occur for some adolescents who are found to be sufficiently competent” (1995:1240). The implication from the report is that there would be few exceptions.

Justification for an exception under these guidelines would depend primarily on an evaluation of competence, voluntariness, and an adequate understanding of information on the part of the adolescent. The authors advise that case-by-case evaluations as to benefits and harms, decision-making capacity, and voluntariness should include several professionals (pediatricians, psychologists, and ethics committees) and not just geneticists.

As cited earlier in this study, Hill and Fortenberry (1992) conducted two surveys to identify the features of images of adolescents. Each survey demonstrated that adults perceive adolescents in negative ways, with medical students appearing to have the most negative perceptions. This research raises the concern that stereotypical images of adolescence prevail in the medical profession and might unfairly bias decisions concerning an adolescent’s competence.

Michie (1996) criticizes those who are against the testing of children, arguing that such positions are based on a fear of consequences for which there is no empirical evidence. She believes that if behavioural or social scientists had been involved with the working party they may not have suggested the same consequences of testing minors that have been evoked by geneticists.
As cited earlier, research on adolescents has shown the benefits of granting decision autonomy. Adolescents who are involved in decision-making and who feel in control of an experience that affects their welfare exhibit increased self-esteem and coping. Clarke and Flinter (1996) in their article against the genetic testing of children admit that “granting choice and control with respect to genetic testing . . . may be of value to the adolescent’s self-esteem and hence his or her coping strategies” (1996:174). Elger and Harding’s research into the hereditary breast cancer gene (BRCA1) led them to argue “respecting adolescents’ autonomy in decisions concerning genetic testing increases their sense of themselves as active and responsible participants in questions related to their own psychological and physical health rather than as powerless victims of adults and genes” (2000:119).

It is my contention that adolescents are individuals with inherent needs and the capacity to come of age in their own right. As stated in the literature review, research on children’s decision-making abilities has provided much evidence that 14-year-old adolescents reason as maturely as adults in medical decisions (Melton, 1993). The problem is how to distinguish mature adolescents capable of making competent decisions from those adolescents who may not be as competent to make decisions about major health care.

There is no agreed upon criteria on which to base maturity and decision-making capacity (Committee on Bioethics, 1995). Drane (1988) has proposed the concept of a “sliding scale” of competency. In the case of very risky or harmful decisions, the threshold for competency should be high. In decisions involving a lesser risk, we can admit a lower threshold for competency. Regardless, the American Academy of Pediatrics states that in many cases the adolescent’s consent or dissent should be binding (Committee on Bioethics, 1995).

Individuals become capable of understanding the future implications of testing and of making an informed decision at widely different ages (Wertz et al., 1994). In this research adolescents began to contemplate their future at the age of fifteen. Although the participants in
this study is small and may not represent the larger population of adolescents at risk for HD, it is my recommendation that health care professionals clearly identify the difference between children and adolescents when considering the risks of predictive testing and to allow access to predictive testing to adolescents 16-years-of-age or older. It should be carried out on an individual basis based not only on the adolescent’s competence but also on informed choice, adolescent’s understanding of HD, illness and the impact of HD in the adolescent’s life and the adolescent’s family, and the support the adolescent perceives in their family and community.

Youth can be seen as acting intentionally – with agency – to find meaning in their lives. It is a process that involves choosing between alternative courses of action. In order to advocate for adolescents, we need to begin to understand how adolescents are active agents engaged in the production and management of meaning in their own social lives. It is important for health care professionals to explore with adolescents what their experience of adolescence is like. It may well be that what is perceived as behavioural disturbances among adolescents are responses to problems within the culture.

As discussed previously, Waxler-Morrison et al. (1990) explains that healthcare professionals and patients can bring different notions to the clinical encounter. Professionals bring the biomedical culture as well as their own ethnic and socio-cultural background. Patients bring their own ideas about the biomedical model and their own set of beliefs and values about illness. There are often discrepancies between these two perspectives in the explanation of disease and illness, the expectations of how each should behave, and of treatment results. The point is that these differences can lead to misunderstanding, and that health professionals must be willing and able to recognize their own assumptions and to open them to reassessment for each health care encounter (Burgess et al, 1998), including those with adolescents. Health care professionals can develop a better sense of adolescent competence by exploring with their adolescent patients salient issues such as beliefs about illness, expectations of treatment, and how
illness is managed in daily life. As demonstrated in this research, identifying adolescent purposes in relation to information about HD (i.e., beliefs about personal susceptibility to onset of Juvenile HD), of requests for predictive testing, or struggles with relationships can establish a therapeutic allegiance that is more relevant and ethically sound than a mere evaluation of competence.

Summary

I believe that this dissertation makes a significant contribution to an understanding of the at-risk adolescent experience of living in a family with HD. The study elucidates the adolescent experience of learning about HD, the impact of this knowledge on relationships with family and friends, and on self-understanding, self-identity, and future self.

An important contribution of this research is the explication of the process of learning about HD that is presented through the experiences of the adolescents themselves. The ability of children to intuit family "secrets" and the impact of these secrets on the children sheds light on the importance of open communication in a family with genetic disease. Parents as well as clinicians have been hesitant to broach the subject of HD with children for fear of inducing negative reactions. This research reveals that children intuit that something is happening in the family, and that it is the uncertainty and secretiveness that creates the stress and anxiety rather than the information about HD. The study also shows that by the time children reach early adolescence they have learned not to communicate about their concerns or fears regarding HD. They learned early in childhood that it was a family rule not to speak about the disease. Consequently, adolescents are often perceived as being comfortable with the information about HD and their risk status when, in fact, they find the information challenging to their sense of well being.

The adolescents in this study did not seek support from their families but were able to find the support they needed through friends and mentors. They engaged in complicated coping
strategies that involved choosing between alternative courses of action, and displayed maturity beyond what would be expected from their age group. They acted intentionally – with agency – to find meaning in their lives as they coped with the challenges of adolescence in general and the additional challenge of being in a family with a genetic disease. This study clearly indicates that healthcare professionals need to re-evaluate their views of adolescent autonomy and capacity for decision-making. It is imperative that adolescents come to be looked on as unique individuals with amazing capacities to handle difficult situations – as such they have much to contribute to further research into inheritable disease.
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APPENDIX I

REQUEST FOR PARTICIPANTS
CONSENT

Your child's participation in this study is entirely voluntary and may withdraw at any time, as well as parents can withdraw a child at any time. Your signature below indicates that you have had your questions and concerns satisfactorily answered and that you are willing to allow your child to participate in the research. This consent form is only a summary of the information and your child's continued participation should be based on satisfactory responses to all of your questions or concerns.

I consent / I do not consent (circle one) to my child's participation in this study.

Please retain the second copy of the consent form for your records.

Parent/Guardian name: ______________________________________

Signature: ___________________________ Date: ______________________

Witness: ____________________________ Date: ______________________
CONSENT

Your participation in this study is entirely voluntary and you may withdraw at any time. Your signature below indicate that you have had your questions and concerns satisfactorily answered and are willing to participate in the research. This consent form is only a summary of the information and your continued participation should be based on satisfactory responses to all of your questions or concerns.

Please retain the second copy of the consent form for your records.

Name:  

Signature: ___________________________ Date: ___________________________

Witness: ___________________________ Date: ___________________________
QUESTIONS/LIFELINE INTERVIEW SCHEDULE

1. Rapport and Information Giving Questions

Do you know who I am and why I am here? (explain if necessary)
Explain what an interviewer is and what kinds of questions will be asked and approximately how long the interview will be. (explain lifeline)
Do you know what I am going to do with the information you give me? (explain and include information regarding confidentiality)
Do you have any questions about the interview and how it is going to be conducted? (comment on the adolescent speaking only about what they want to, and remind the adolescent that they can stop talking whenever they feel uncomfortable)

2. Lifeline

In order to sensitize and assist the adolescent in accurately recalling her/his experience, the adolescent be assisted in filling out a “lifeline.” The lifeline will be constructed by drawing a line across a blank sheet of paper. The beginning will be marked by the adolescent’s first knowledge of HD in the family and by the learning of her/his at-risk status for HD; the end of the lifeline will be indicated by the present time of the interview. Subjective and objective events and experiences will then be marked at different points along the continuum, in chronological order. Finally the adolescent will be asked to tell their story of living with HD and at risk for HD using the lifeline as a guide. At the end of telling the story of living with HD, the researcher will ask the adolescent a final question regarding the potential impact, if any, of the adolescent’s at-risk status on her/his view of the future.
The role of the interviewer will be to facilitate the telling of the story through active listening, reflection, empathy, and some probing to ensure that a coherent and complete story of the adolescent’s experience will be obtained.

3. Closing

Do you have anything more you would like to tell me that I might have not asked you?
Do you have any questions you would like to ask me?

The interviewer will state that talking about HD and being at risk for HD could potentially bring up feelings that might be uncomfortable, and, therefore, will encourage the participant to contact the interviewer if intrusive or upsetting feelings or thoughts arise.
APPENDIX IV

ADOLESCENTS' LIFELINES AND STORIES
Stephanie: Age 14

- Knew something was wrong
- Two incidents of dad being violent with mom, grandmother, and me
- Attend monthly adolescent-at-risk meetings with HD society
- Dad hospitalized for two weeks and put on medication
- Dad's still calm but I still don't trust him
- Getting more in trouble with Mom

AGE | 8 | 9 | 10 | 11 | 12 | 13 | 14

- Dad had predictive testing
- Mom brings big book on HD
- Told HD is hereditary
- Mom put me on waiting list for psychiatrist
- See Dad's movements
- Incident at lake with Dad and Grandmother
- Friends notice Dad is different
- Dad lost licence
- Dad given more medication to calm down
- Told friends about Dad's HD. Now all friends know
- Attend national HD conference in Saskatoon
Stephanie was the first adolescent to volunteer to participate in the study. We had met at a group session for adolescents at risk that I conducted for the HD Society of BC. I was initially concerned that Stephanie’s mother’s enthusiasm about the research was based on her belief that there was a therapeutic benefit to be derived from the interview process, and that Stephanie may not have wanted to engage in the research but was doing so to appease her mother.

I met with Stephanie and her mother in the family home and voiced my concerns to both of them. I was assured by each of them that they knew the parameters of the study and that Stephanie wanted to tell her story. I also was able to meet Stephanie’s father, who only said hello and then continued to watch television. Stephanie did not feel comfortable talking at home and wanted to be interviewed at a sushi restaurant ten minutes from her home.

I knew that something was wrong. My dad was different.”

Stephanie knew “something was wrong” with her dad when she was eight years old. “He would act weird.” She would go to friends’ houses and notice a difference between her father and her friends’ fathers. She would think, “Why did my dad act like that? What did he do that for?” She never asked her mom about it because she thought that must be “the way it is.” When she was growing up, she knew something was wrong with her grandfather by the way he moved his feet. She didn’t know that it was Huntington’s Disease. It was only after her mother brought out a big binder with information on HD that she began to understand that the differences she noticed in her family were symptoms of HD.

Stephanie’s aunt came to visit and Stephanie realized she had HD. “I can tell. She’s like kind of moving and her hands are going and feet kind of.” Her mom told her later that her aunt had HD. That was a validation for Stephanie; she realized that what she perceived as symptoms
of HD were accurate. Stephanie then became privileged to many “family secrets” through her mother about her father and her father’s family. For instance, she knew what was put on her grandfather’s death notice, the status of a missing uncle, and the family’s belief about who had HD and who didn’t.

She also began to emulate both the verbal and nonverbal communication that her mother used when she talked to her father or reacted to relatives.

It was really funny because she looked at my mom and she had, like, the puckered mouth, and my mom kind of sat there and I looked at my mom and I looked at her and we started laughing. And my aunt was just sitting there and it was so funny.

During an argument when her father became violent she recalled responding to her father in the same way her mother would respond.

He walked over to me and I said, what are you doing? You should be downstairs! He started yelling. And so I looked at him and I pushed him.

Stephanie has a strong alliance with her mother and speaks of her father and her father’s family using the term “we” instead of “I” in most of the conversation.

We don’t visit my dad’s side if he’s not there. We really don’t like them. They’re really conceited and think they’re everything. Always want to know what we’re doing. How much money we’re spending.

When Stephanie was eleven her mother brought out a large binder full of information about HD. Stephanie didn’t want to listen to the information. She listened for a little while and then went to bed. She said she wasn’t scared of the information because she felt there was “going to [be] a cure in, like, two years.” [The information concerning a cure was told to her after her mother listened to a TV interview with a geneticist.] She saw HD as something “so far away.”

Stephanie doesn’t have a close relationship with her father. “He wakes up at nine, has his breakfast and goes downstairs and watches TV until nine at night.” When asked what her father knows about her she replied,
[He knows] that I play hockey and stuff like that. I do modeling now. He didn't know before and my mom didn't want to tell him. He worries about money.

She learned this from her mom. "I just kind of end up learning from other people. I'm always finding out stuff." Stephanie says that even though she doesn't do much with her dad she is not missing out on anything because "my mom is the Mom and Dad." Stephanie also stated that "we [Stephanie and her mother] don't talk about that many things." Personal subjects are discussed only when Stephanie brings them up. Stephanie doesn't like to talk with her mom or be seen with her mother even though she recognizes that her friends feel ok about being with their moms.

Stephanie's Mom arranged an appointment for Stephanie with a psychiatrist.

She thought it [would be] good. So that I had something to lean back on just in case like I ended up freaking out or wanting to [ask] questions.

Shortly after that time, Stephanie had an angry encounter with her dad when he would not return home from a visit at a local lake. He also had a car accident with her in the car that resulted in his license being revoked. Her mother phoned the DMV to have his license taken away. She described his reaction to his license being revoked as, "He's a baby. That's why he lost his license. He's a dummy." At the same time, Stephanie felt that the removal of his license made HD real. "He had his license taken away and then it was real." It was also during this time that her friends began to recognize that something was different with her dad.

There were two physically aggressive incidents that occurred with Stephanie's father during her twelfth year. The first incident happened when Stephanie and her parents were making a cat scratching post. She was angry about her father's comments. "I got really mad and took a piece of wood and hit it on the seat cause he was just, don't do it! Don't do it that way. No! No!" Her father got angry and picked up his cane and hit her on the back. "It hurt but I wouldn't, like – no tears came."
The second incident happened when Stephanie, her mother, and grandmother were sewing “Quillows.” Stephanie believes it was the result of her father being left alone too long downstairs. He knocked something off a sewing machine with his cane.

My mom says, oh, good job, you’ve broken it. And he got all mad, and he starts looking at me. Like to talk to me or whatever else. It’s just like, oh, my God. Like my grandma, she just looked at him, she’s like, you stay away from her!

He knocked Stephanie’s grandmother over.

She had fallen really, really badly. She had bruises all over her arms and [he was] grabbing her and throwing her and everything . . . and then he came at me and he was about to hit me and I pushed him and he hit me across my back. My mom got involved then and she was like, you don’t push my daughter! You don’t hit my daughter! Go to bed right now!

He hit his wife on the hand and the Stephanie said that they all started yelling at him. Stephanie was scared “cause that hurt like twice as hard as the time before.”

Six months later her father went into hospital for assessment and medication. Stephanie said that her father started yelling.

He just sat there and made us do stuff. It’s like we’re not going to put up with this anymore. And then we just kinda sent him to the hospital.

Although Stephanie used the word “we” in describing why her father was hospitalized, she did not know that her mother was taking him to the hospital.

She came to pick me up and she said, oh, we have to go to the hospital. I was like, why? And she’s like, oh, because I put Dad in there. He’s acting up again. He wouldn’t take his pills. He made my mom get his pills.

When he returned from the hospital about two weeks later he was taking his medication. It took time for Stephanie to feel safe with her father. Stephanie says that she does not trust her father since the violent incidents.
When he did yell, when he did hit me...I hated him. But now, nobody dislikes him. He won’t bother anyone pretty much trying to talk to him. But he won’t talk back. It’s very weird. Its like you are talking to a brick wall. He just sits there staring at the table. Its like, ok, thanks for listening.

At thirteen Stephanie told all her friends that her father had Huntington’s Disease. She attended an adolescent-at-risk support group through the HD Society and began to attend National HD Conferences. With regard to her future, she felt Huntington’s Disease wouldn’t change her life; if she had the gene she would just live her life “faster.” She believes that there will be a cure soon, but if she has the gene she would have a life that is different from her father’s as well as her mother’s.
Carla: Age 15

- Knew something was wrong – Mom yelling
- Mom chasing Dad with knife
- Parents separated
- Sent to psychiatrist
- Mom placed in care home
- Took tour of genetics centre and learned new information about HD

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- Grandfather in hospital with HD
- Told about HD and that it was hereditary
- Mom threatens me with bowl and knife
- Understood what hereditary meant
- Visit Mom every Monday at care home
2. Carla (Age 15, only child, Mom has HD, Dad has a partner with a 14-year-old daughter)

I had known Carla since she was eight years old. Her mother was the first client I worked with who had Huntington's Disease. It was through my relationship with her mother that I became involved with the HD Society and with predictive testing. Since Carla's mother had been my client for several years, I was uncertain how that might affect the interview and the questions that I would ask her. I was privy to a great deal of knowledge about the family history, and I wondered if Carla would recount incidents that her parents had shared with me at an earlier time.

I met with Carla at her home, where she lived with her father and her father's partner and child. I voiced my concerns to her father, his partner, and Carla and was assured that they felt my knowledge of the family would be helpful and not a hindrance. Carla's mother had been placed in an extended care facility earlier in the year and, in fact, Carla had chosen a night for the interview in which she had spent time with her mother at the care facility. We held the interview in a basement daycare facility attached to the home. Carla's father, who appeared nervous about the emotional well-being of his daughter, regularly interrupted us with pizza.

"I see somebody I don't know but who I've lived with my whole life."

Carla remembers that when she was five her mother was yelling at her father a lot. "My dad telling me there was something wrong. I always figured something's wrong in our family. Sometimes I thought it was my fault." As she grew older she remembers a lot of "violence" in the household. "All I knew was, my mom was mad at my dad, like, every single day." When she was nine her mom chased her dad around the house with a knife. "She locked herself in the bathroom and we could hear all these crashes. We thought she was going to commit suicide but she came out after and she had pulled down the towel rack and busted the shower." When Carla was thirteen her mother "chased me around her apartment with a knife and threatened to shove a
bowl down my throat.” She found that whenever her mother was mad she would threaten her. However, Carla often contradicted herself in the conversation. When asked if her mother was always mad at her she stated, “She would rarely yell at me. She never really put anything on me.” At times she would speak of being scared but she felt that, “She’s my mom. And I am not going to just give up on her.” Carla was very upset that her mom’s friends “gave up on her.”

Carla was six when she first heard about Huntington’s Disease after visiting her grandfather in the hospital. Before her grandfather entered hospital “he would sit in one chair more, and he could talk less and he could walk less so I knew there was something wrong, child intuition or something.” She remembers him fondly for playing dolls with her in the hospital and letting her wheel him in the wheelchair.

When she was ten her father talked to her about HD and informed her that it was hereditary. The family had attended a predictive testing session with Carla. All she remembers is wanting to know if her mother was “going to die tomorrow” and that she “had no idea what to ask.”

I walked away thinking that my mom was dying and I had no idea when. And I didn’t care that I had it, that I had a chance. I was just like no, my mom can’t leave me.

When asked if she still feels that way Carla replied, “I never really knew my mom, which is quite sad. My dad always talks about her, how she used to dance, and how she used to be so good. I would give anything just to see that. I think back about how I don’t know my mom, yeah. And there’s nothing I can do about it. . . . [Now] I see a disease. I see somebody I don’t know but who I’ve lived with my whole life.”

Carla remembers very little about the time she was eleven. It was during this time that her parents separated.

I just remember the breakup. The breakup just killed me. I think I went into a kind of depression because I was like, why is this happening? Is it happening because
of me? Is it happening because of the disease? Is it happening because my parents don’t love each other any more? Is the family falling apart?

She was seeing a psychiatrist during this time. When asked about whether she wondered which parent she would live with she stated, “I knew enough to know I couldn’t really live with my mom. That she couldn’t take care of me as well as my dad could. A lot of times I was worried. I mean I love my mom, but I knew she couldn’t take care of me.”

When Carla was fourteen she attended a tour at the Centre for Molecular Medicine and Therapeutics (CMMT) with the HD Society. The tour explained HD in detail, and included several question and answer periods. “I knew that HD was hereditary but I learned the most about it on the tour.” During this time Carla’s mother entered a care facility. “Even though I knew it was a deadly disease, I always kind of had that hope that she’d get better but when she went into the care home it was definitely like, that’s it, there’s no hope.

Carla speaks of her relationship with her mother in contradictory ways.

My mom was there, always, but . . . her sad days, it was like she was never there, really, unless there was something that I was sad about. If she was fighting she didn’t want me to know about it but of course I’m going to know. I can hear her. But I guess she tried to protect me by not talking about it.

At age eleven one of Carla’s biggest problems was reconciling her father’s new relationship while dealing with her parent’s separation. Carla kept the relationship a secret from her mother. Her mother did eventually find out when Carla was twelve, when she and her father moved in with her father’s partner. “I have the feeling I’m just going to have to live with it.” Carla told the psychiatrist nothing about this. Carla has anger toward her father because she feels she is always the last to know what is going on with her mother. Each major event on the lifeline is followed with a comment about who knew what was going on before her. This was particularly evident when her mother moved into a care home.
I didn’t know until a couple of days later. This time, many more people knew before me. It was really bad. I mean I had friends who knew about it before me. I couldn’t believe why he would keep something like that from me. I mean for the first time I just started crying.

When asked about times that are good with her mother, “There were a lot of happy times as well. I mean no full life is full of sorrow.” Carla remembers times when her mother was in hospital and still could comfort her. She states there are times when, “You can see her actual personality. You don’t see the disease. You see her, which is nice.” She describe her mother as “a really, really nice person that really cares about other people – somebody who . . . puts their life on the line for you.”

Carla visits her mother at the care home every Monday. She feels more comfortable with her mother there “because she has people looking after her 24/7.” Carla is not as concerned about her mother dying as she was in the past.

When I was younger I used to think it was like in the next week. As I grew up, I started thinking, like, oh yeah, maybe in two years max. Like that’s what she has. But now, it could be years. It could be days. It could be months. I don’t know.

Carla would like to be tested now.

I could have this great career in singing that could be stopped by something that I can’t control. I’d try and get it [career] anyway, but I think it’s better to know. Because then you can plan out your life into a kind of compact space.

When asked about her future she replied,

I look at my future as obviously me becoming a singer. It may become something having a huge career. But I also see another path that might happen. And I watch my mom and I see me in the future and that scares me. A lot.

As Carla began to explore her future she acknowledged that she had been depressed since she turned thirteen. She did not talk with her father about it because “he’s got enough on his mind . . . we’ve both got pretty hard lives.” She also does not want to go back to the psychiatrist
because she is “reluctant to speak to people I don’t know.” She has good friends who “really care” and finds that helpful.

I feel like my whole life I’ve never really felt love. I mean I’ve felt love, but I don’t think I’ve felt enough love that an only child should. I have an empty feeling.

When asked if she ever thought about taking her life she spoke of a time when she considered jumping out of her bedroom window.

All I have to do is climb to the ledge and jump. That’s all I got to do. I may not kill myself, but at least I’ll be injured. I do feel some of these things are my fault. I feel like I should be punished. There were just so many times when I thought about taking my life.

Carla “loves” the Spice Girls and looks up to them as she couldn’t to her mother.

All my life I figured that little girls should look up to their moms. Girls are supposed to look up to their moms. Boys are supposed to look up to their dads. I should look up to my mom. And I never had a chance to look up to my mom.

She described her relationship with Melsie of the Spice Girls.

I never thought you could love someone so much without knowing them. But when I look at Melsie . . . it’s so weird. I mean I would look up to her more than I would look up to my mom.

Carla ended her interview by singing a song she wrote her mother called “Sing with You Again.” It was in this interview that I was grateful for being a therapist. I experienced Carla as an adolescent at risk for more than HD. She was a traumatized young girl who suffered from depression and tried desperately to make sense of her life by taking responsibility for her mother’s illness and well-being. She engaged in suicidal ideation, and at one point in the interview process I engaged in a structured debriefing process to help Carla defuse some of the intense emotional reactions she experienced from past situations of trauma.
I was concerned about the interview creating more distress for her, but in follow up I discovered that Carla felt a therapeutic benefit from sharing her feelings with me. She felt that she had never been able to tell how she really felt about her life and the process gave her a chance to begin to make sense of her feelings.
Marilyn: Age 16

- Told Grandfather is sick in hospital – he was scary
- Aunt was moving funny

- Started listening a lot more to family talk
- Aunt lost job and had accidents with car
- HD still just a name

- Friends thought Dad was gay
- Embarrassed
- Associated Dad’s behaviour with aunt’s movements
- Video of Dad climbing pyramids – we all saw HD movements

AGE  7  8  9  10  11  12  13  14  15  16

- Grandfather died
- Told aunt had HD
- Sunday dinners felt bad – didn’t want to be by aunt

- Summer vacation, Dad depressed, I figure out he has HD
- Told brother I thought Dad had HD

- Aunt told me Dad had HD – she thought family had told me
- Told about HD and its hereditary nature day before 16th birthday
- Brother learned of HD
- Grandmother would not believe Dad had HD
- Everybody feeling sorry for me
- Dad quits work
- Everything changed – I really hate him
3. Marilyn: (Age 16, has a younger brother, Dad has HD)

Again, I found myself having a great deal of prior knowledge about the Marilyn’s family and their concerns for her. I spoke to Marilyn and her parents about these concerns. Marilyn wanted to tell her story as long as she could be assured that she would be able to speak to me alone. We conducted the interview at my office outside of office hours.

“When you find out it is true, it’s almost like your mad not sad.”

Marilyn related two significant memories from when she was seven years old: her grandfather in the hospital and her aunt’s presence at Sunday dinners. The memories of her grandfather are of him in a room with a “smell of sickness and dying.” She found her grandfather “scary, he looked like a monster.” One of her concerns was that the sickness was contagious; she did not talk about that with anyone.

Her aunt would attend Sunday night dinners and sit next to Marilyn. Over time Marilyn noticed that she “ate funny and moved funny.” She did not think of her aunt having symptoms, but thought that her behaviour was just “dumb.” At age ten her grandfather died and she was told that he had Huntington’s Disease. During that same year she found out that her aunt had HD. “I kind of clicked the two together. I thought well, no. Because every time I kind of clicked two things, I would deny it.” Marilyn described her family as “secretive” and would not talk to anyone because “it is kinda uncomfortable talking about it in front of them because we’re so secretive.” The one person Marilyn felt comfortable talking to was her mother. “She is pretty much the only one in the family that really isn’t that way.”

When Marilyn was twelve she “observed a lot more things.” She entered high school and was starting to make new friends. “They kind of thought my dad was weird.” She says that now when people meet her dad they ask why he has such a high voice. “It’s just like, shut up. I’m sick
of it.” Marilyn says that she will tell people that he is dying from a disease. “That shuts them up.” She feels that it is preferable to let them know that he has a disease he is dying from than to have people think he is “gay.”

Marilyn did not know if her father had had predictive testing but assumes he did because he “found out in 1996.” Although Marilyn had been given information about the disease, she was confused about whether she was 25% risk or 50% risk. During the interview we were able to clear up her confusion, and she found it helpful to have correct information. It was at Christmas that Marilyn “put everything together.” She remembers her aunt almost in tears after the family talked with her. “I thought something’s up. I asked my mom. She told me. She’s the only one that tells me anything.” Marilyn feels that she has to figure out what the question is before she can get an answer. “It’s kind of like I’m a detective.”

In the spring of Marilyn’s twelfth year the family traveled to Ixtamico, Mexico. They visited a famous temple, and Marilyn and her father climbed the stairs to the top of the temple while her mother captured the trek on a video camera. When the family returned home her father watched the video.

That’s when he kind of saw himself, you know, with it. And he thought, oh, my God. You know. I might have been tired but I couldn’t have been that tired. So, that’s why they got testing.

In the summer of 1996 the family vacationed in a remote resort. Marilyn noticed that her father was behaving differently. “He would just sit there, you know, and look like he was about to die. He would not talk.” She realized her dad was depressed and began to try and figure out what the problem was. Her mother told her that “he was going through a really hard time.”

I thought, a hard time? What is he going through? You know. And I was kind of trying to picture it. Was it work? No, it was not work. So, I don’t know why I guessed it but I just guessed. And then I thought that’s what it is! That’s it!
She told her younger brother but he disagreed with her. When asked why she didn’t ask her mother she replied, “I guess I really didn’t want to know.”

When Marilyn was fifteen she realized that her friends thought that there was something wrong with her father. “Why does your Dad have such a high pitched voice? He sounds like your brother. Oh, he walks funny.” Marilyn felt that they didn’t “mean anything by it.” She became increasingly uncomfortable being with her father in public. “Like, if he comes for parent teacher interviews with my mom, it’s like, oh God – don’t!” She explained that she did not want attention drawn to her by her father’s conduct. She felt that he has always been “kind of annoying but now with HD, he’s really annoying.” Her way of coping with her father’s behaviour is to “avoid contact.” “I try to avoid any contact with other people and him. Yeah. That’s why I never bring friends over anymore. It’s just too awkward because he asks them all these questions. He’ll ask them the same questions like five times!” When asked if she thinks his persistent questions are associated with HD, she replied, “I don’t know.”

Close to Marilyn’s sixteenth birthday she noticed that her aunts suddenly started to show an interest in her. “Not interest but, Oh, how are you?” Her aunts had never spent a great deal of time with her yet now she was invited to go shopping for “anything you want.” When Marilyn finally asked one of her aunts about all the attention she was told, “I am so sorry about your Dad.” Marilyn reacted with shock and it was then that her aunt realized that her parents had not told her that her father had HD.

Shortly after the shopping trip with her aunt, Marilyn spent a day shopping with her mother. “I was pretty mad all day. I wasn’t pissed off with her but with the fact that it might be true.” After she returned home her mother asked her what was wrong. Marilyn blurted out, “You know, Daddy has Huntington’s.” Her mother started crying. “Oh, no. Is that what you thought? Oh, Marilyn, I’m so sorry. . . . I’m like, no. You’re supposed to say, No, he doesn’t have it!”

Marilyn spoke about how her parents began crying and yet she had no tears.
I didn’t know how to react to it. My whole life I knew it but I didn’t want to think it and now I knew it. I knew it was true and that was hard to deal with.... I didn’t know what to say. It was really so awkward. I was just like, ah...because you always think it won’t be true. And when you find out it is true, it’s almost like your mad not sad. I was really mad at myself for not thinking that he had it. If I had only not asked. Now that I have the evidence knowing that it was true, like, I don’t want it.

The events with her aunt and parents occurred a few days before Marilyn’s sixteenth birthday. It wasn’t until several days later that Marilyn talked with her mother and father. When Marilyn questioned her parents about why they had kept this secret her mother replied, “We wanted you to get through your life without knowing.” Marilyn could not believe that her parents thought she was “stupid” and couldn’t figure it out. It was at this time that the hereditary nature of Huntington’s Disease was explained in more detail. “When I heard that you have a 50:50 chance, what I heard was it’s either Thomas or me.” After it was explained again Marilyn asked, “You mean we can both get it! But you mean we can both not get it, too?” Marilyn had never thought about heredity in those terms.

Marilyn believes another one of her aunts has HD.

I’m pretty sure she has it. It doesn’t affect me as much now. It’s kind of like – ok, it’s another one. You know three out of five people have it. And I know she has it. Just because I know.

Marilyn talked about her father since he quit his job.

He’s at home pretty much every single minute I’m there. It’s pretty hard because I’ve never really liked him...I’ve just never had a good relationship with my dad. So, now he’s changed so much that it is almost like living with another man.

She talked about how he does not help her mother around the house because he feels he “can’t do it.” She believes that he uses the disease “to his advantage,” which contributes to her dislike for her father. Marilyn fights a great deal with her father, which results in “frustration” for her mother.
She's mad at me. She says I push my dad over the top. In a way I guess I do. It's my therapy for proving – see, see he's a monster. I don't mean to, but in a way that's how I prove that he is not normal.

Marilyn believes that her father still has a choice in his behaviour, and when she pushes him she can see the disease and realize that he can't "put the effort in to being the father that we need." When asked about the last time she was with her father "without the disease" she spoke of a time when she was eleven years old and he asked her to go roller blading in Stanley Park.

When asked about her future Marilyn talked about how there might be a cure when she is at the age of becoming symptomatic. Marilyn is ambivalent at this point as to whether she wants to take the predictive testing.

If I do get it, I don't really want it... to live to that stage where I am in a nursing home drooling out of the side of my mouth, getting my Jell-O... I would rather live my life to the fullest and then die. I guess I associate the end with my grandfather because that's what I saw.

Marilyn feels that because she has seen how Huntington's Disease can change people's lives she is going to live her life more fully.

If I establish my life... now then it won't really matter because I'll have everything that I have dreamed and strived for before it ends. You know. Don't have any regrets is basically my future. Live my life the way I want to now.

Throughout the interview I was struck by the anger and frustration that Marilyn experienced in her life. She spoke in angry tones and gestures about her father. At one point in the interview when she was recounting an incident with her father in a public situation, she seemed filled with contempt. I became concerned that she might lose control of her feelings in a destructive way.

I have seen Marilyn on several occasions since the interview and have become actively engaged in helping the family deal with the consequences of her father's unemployment and the effects of his behaviour on Marilyn and the family.
John: Age 18

- Meeting with psychiatrist and family – told Dad had HD
- Mom wanted me to talk about my feelings – I didn’t talk
- Saw Dad “twitching”
- Talked to Mom about my feelings
- Moved to new school
- Dad lost driver’s licence
- Dad got depressed and stopped going outside apartment
- Lost respect for Dad

AGE | 11 | 12 | 13 | 14 | 15 | 16 | 17 | 18
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- Felt depressed about a lot of things
- Felt pressure: If Dad got it at 35, what about me?
- Moved to new town with Mom and sister

- End of grade 10, Dad moved to same town
- Saw Dad a lot
- Went on Internet to get information
- Friends found out Dad had HD

- Met Dad’s friends with HD
- Reduced my visits to Dad to once every two months
- Decided to get PT for self
4. John: (Age 18, older sister, Dad has HD)

Again I found myself in the position of knowing a great deal about the family before I met with the adolescent. John’s father had attended the HD summer camp where I was a volunteer, and I had had many experiences with him. He was a man prone to anger to inappropriate displays when he was agitated. I had worried about his capacity to lose control at camp and had been part of a team decision to remove him from camp and send him home early.

John did not know of these incidents but did know that I had met his father at camp. He assured me that he felt the interview would be helpful for him to get his thoughts together, and he hoped that other teens would benefit from hearing his story.

We met at the home he shares with his mother when she was at work. The interview was occasionally interrupted by a number of friends who were engaged in a study group downstairs. Although this seemed to be disruptive at times, John felt that the fact he was being interviewed demonstrated to his friends the significance of his father’s disease and it’s impact on John.

“My dad’s not drunk. He has Huntington’s.”

John was twelve when he realized that “something was wrong” in his family. He stopped seeing his father regularly, and his parents were not talking to each other. Shortly after that time, John’s mother took John and his sister to a meeting with a geneticist, a psychiatrist, his mom’s best friend, his Dad and his stepmother. It was there that he first learned about Huntington’s Disease, as he had no contact with any relatives that may have been symptomatic. “If there were any signs from Huntington’s, I said it was the accident” [his Dad’s motorcycle accident]. At the meeting his father read a letter to the children “to express his feelings.” It explained that he had wanted to let them know right away about his illness but John’s mother had not wanted the kids to know.
My mom was pretty angry and hurt and so was everyone else. I was more like this is about my dad having Huntington’s, and so it kind of went bad from there.

When asked what he learned from the meeting, John commented, “I learned that he was not going to die right away. It usually takes a while to set on, and I have a chance of getting it.”

His mother wanted to talk about the meeting but John “wasn’t ready to . . . for a little while.”

I don’t think it actually hit me that it could really affect me. I was just kind of worried about my dad – that he was going to die pretty soon. I still had the image that it was going to be a lot quicker than what they told me. So, I started talking to my mom I think around thirteen when I actually started noticing twitching.

John and his sister talked after the meeting and both decided to throw away their copies of the letter. When asked if he and his sister talked about their risk status John stated that, “we didn’t say much; I don’t think either of us ever talked about that.” In retrospect, John felt that his parents’ fighting and the conflict in the psychiatrist’s office affected him more than the disclosure of HD. “I was pretty stressed out already as a kid. I wanted to kind of forget about it [HD]. There was one too many things to think about in my life.”

John and his sister continued to see their dad on a regular basis.

I think I did start to notice some changes. I didn’t know it was from the disease. But we did start to notice. Like sometimes he would flip out on us. You know. It would be awful. He would suck and blow and suck and blow. And then he would come back and be fine.

He said that he and his sister were not afraid of their dad’s reactions, “When he spa zed you knew he was mad but you just couldn’t figure out why he was mad but then you just gave him a break.”

John’s mother moved the children from the East Coast to Victoria when John was fourteen. John described the move.

It was really rough because I was kind of depressed at that point. The Huntington’s sort of affected me. I started thinking . . . how quickly it came on to my dad. My dad was like thirty-five. Christ, so I have that long before I start like
shaking and things, so I better hurry up. I started feeling a little pressured that way – that I want to do all the things that I want to do. The first year was hell.

He felt his depression was not only from learning about Huntington’s Disease but it was a combination of things. “I didn’t fit in. I am tall. I stand out. I was being heckled. I couldn’t take a joke. I could get violent that way.” John felt the move was “really hard” on his sister. “She had a lot of friends when she was in grade nine. She brewed the whole way across and she was crying. She was pretty resentful for a while.” He felt calm because he “respected” his mother’s decision.

At fifteen John decided that was “tired” of the depression and “realized that I got to get out of it.” He made some friends, moved to a new school and decided “to change.”

So, I cut off all my hair. I changed my name from Johnny to John. The one good thing I liked when we moved from Toronto to here was that I didn’t have the same reputation. So, it allowed me to start again.

John did add, “You know I am still not quite over it but at least I’m getting a lot out of life.” It was shortly after this shift that John’s father moved to the West Coast and he began to see him regularly.

It was during this time that John began to disclose to his friends that his dad had a disease.

I said it was kind of like Parkinson’s because people know what Parkinson’s is but a lot of people don’t know what Huntington’s is. I said it was a disease that made you have muscle spasms and it eventually affects your brain and slowly kills you.

When asked if his friends questioned whether John would get the disease he stated, “Yeah. A couple of them asked. I said, yeah. I could get that. It was a very hard thing to say.” He recounted the time when his friends first met his father.
We were at a birthday party together and we were going to a movie and we saw him walking down the street. And I gave him a big hug and my friends went, Is that your Dad? They thought it was some drunken guy that was walking. Yeah. My dad’s not drunk. He has Huntington’s.

John began to notice more physical changes in his father’s movements. “I was noticing because I know him as well as anyone. I definitely know what to look for because I knew what it was – twitching and all that stuff.” John wanted to learn more; being a computer science student he went on the Internet.

The main thing I learned from the Internet was data. The fact that there has been lots of research done with it. Things that I never had heard about were on the net.

When asked if he found information on predictive testing he said that he had and that it “pretty much” started him thinking about getting tested.

I think my initial reaction was no. It was like, if I don’t know then I can ignore it and go on with my life. And then I came to the realization that it was probably more detrimental to me . . . I am pessimistic . . . I will always be thinking of having it.

John felt that if he tested positive for the gene it would help delay onset of the disease.

I could do all this preventative stuff. Then I’d have ten more years doing more things I have to do then just forgetting about it and missing those ten years.

Yet when queried about the information on prevention he stated that he “didn’t find much.” “I’m thinking that there is going to be a computer program – you know – that I could use.”

John began to feel uncomfortable with his father’s driving. He described his father’s movements with his feet on the gas pedal as unsafe. He did not directly talk to his father about this but decided to “ride my bike all the time.”

So, I would like – I was always avoiding him. I would avoid [a] confrontation kind of thing. I’ve always been a coward with my Dad.
He began to visit his father less frequently.

I didn’t want to hang out with him as much anymore. I lost some respect for him. . . I’ve known my dad forever and he has never had a job. I started losing respect for him for that reason.

John began to see that his father had “bouts of depression.”

He would stay inside and be depressed and then there were times when he was active. When he actually lost his license . . . he didn’t go out anywhere. I felt like he was a totally lost person. . . . I tried to make the effort to see him – so he would remember who I was. I tried but, no, he was just, like, shut down.

When asked if there were any similarities in the depression he experienced and how he saw his father’s depression he replied,

I didn’t want to be around people a lot. I felt as though it was mainly tough work. I don’t know for him. It could have been the same because he ignored people. . . . For me I know I was like trying to do something, and every once in a while I would break out of it and go do something. . . . My dad never did that.

John’s grandmother moved in with John’s father. He feels that it’s a “good thing because they seem to get along a lot better then they used to.”

John has set a date for predictive testing. Prior to arranging for the tests he spoke to his parents. His father told him that he shouldn’t be tested. “He said it was better if you don’t cause then you will go on with your life and you will find things a lot easier. And then he just kind of got quiet.” John’s mother thought he should be tested. ”She thinks that it’s the right decision for me and that I can handle it. They both have to come to the pre-test to see if I am stable enough to take it – enough to do it.” He is uncertain how his sister feels about his decision to be tested. “She thought kind of like it was my reason and that was good, but she didn’t say anything about herself. I don’t know. I don’t know if she thinks it is a good idea.” When asked who he might share the results with he replied, “Well, if I don’t have it then I am telling the world! But if I do have it, I’ll probably tell everyone. I probably will. All the people who need to know.”
When asked about his future John spoke of aspirations of becoming a computer scientist and a traveler. He dreams of going to Japan, and he has spent three years learning Japanese.

I started leaning it in school, but I want to get a degree in computer science, make money, and then go see all the places I want to go see. There are lots of things I want to see, right, and it’s not going to matter to me [whether he has gene or not]. I am going to see them anyway. It is just going to mean how much time I have. It just condenses it.

Since this interview John did engage in predictive testing. I have visited both John and his mother since the testing. John continues to keep in touch with me through email and is preparing for university in the fall.
Daphne: Age 19

- Knew something was wrong – Mom would not let me see Dad
- Moved to new town with Mom and brother
- Nana moved in with Dad
- Moved back with Dad and Nana
- Brother told me he is going to get tested
- Moved in with Dad
- Dad lost licence
- Dad stopped using joystick on computer
- Saw own mortality in Dad’s HD
- Went to HD Society dinner – met woman who had kids my age

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- Meeting with psychiatrist and family – told Dad had HD
- Noticed Dad’s symptoms – poor memory and he dropped things
- Dad separated from stepmom
- Had to move out due to finances
- Girlfriend died
- Major depression
- Stopped seeing Dad
- Suicidal
- Placed in Psych Unit
5. Daphne: (Age 19, younger brother, Dad has HD)

I had some concerns when I interviewed Daphne. She had just moved back with her father and grandmother and seemed to be in a state of confusion. The only place that she was comfortable talking was on her balcony, which was crowded with boxes and furniture. We managed to create a small space where we could place the tape recorder and an ashtray for Daphne. She displayed difficulty in focusing on the topic and would often tell many stories within one story. At the same time, noises from a train yard near her home would challenge my ability to hear her.

As the interview continued, I discovered that she had a history of suicidal ideation and had attempted to take her life. Upon further exploration she revealed that she still felt depressed and knew that she needed help to deal with her state of mind.

“It is like having your mortality shoved in your face every day.”

Daphne was thirteen when she knew that something was wrong in her family because her mother had not let Daphne and her brother see their father “for like a year.” She recounted a meeting that she attended with her father, stepmother, mother, brother, mother’s best friend, a psychologist, and a geneticist.

Mom was there and Dad actually told us. Dad wrote a letter about how it affected him – not being able to see us because of this disease, and how much it hurt him and all this kind of stuff, too, which was valid.

Daphne learned at the meeting that her father had wanted to tell her brother and herself about having Huntington’s Disease but her mother did not want him too. “He blamed my mom and it is not her fault.”

Daphne felt the meeting was a hard place to learn about her father’s illness.
I would have rather known right up front. She’s always protected us but I have always hated it because I don’t feel I need to be protected. I would rather have had the opportunity to do it myself and to seek out help if I needed it or not.

The meeting was difficult. “It was like all this family stuff on top of it. Like no one can understand the family dynamics. We’re all crazy.” When asked what she learned about Huntington’s from the meeting she stated, “Well, it was that I have a good chance of getting it and that my dad is going to die from it.”

Daphne and her brother began to see their father on a regular basis after the meeting. It was then that Daphne started to see some of the symptoms of HD.

I noticed his movements. Like he was pretty shaky sometimes and just dropping shaky sometimes and just dropping things that he would not normally drop. Or his memory; his short-term memory got a lot worse and I had to remind him of things.

When Daphne was fifteen her mother moved the family to the West Coast. Daphne reports that after the move her life went “down hill.”

My mom promised that if I hated it here she would let me go home. And in a year I said that I still hated it here and I wanted to go home and she would not let me.

Daphne remembers her father moving out to the West Coast within a year, and that regular visitation occurred with him after he arrived. The visits ended when a fight developed over child support.

Mom was mad because Dad wouldn’t pay for anything for John. And there was a really long fight, and she got me to go over and tell him that we didn’t want to see him anymore. I was picked to talk with him all the time. And I really thought it was unfair that I was always the one that had to go over and say how we all felt. That really sucked for me because I was always the bad guy.

As Daphne’s story unfolded it became clear that she was under a great deal of psychological distress. As questions evolved that explored the nature of her distress, Daphne
acknowledged that she had “cut” her arms and her legs “and such.” She recounted what had happened to her during her sixteenth year.

I ran away from home because my mom said that I wasn’t allowed to leave the house because she found out about my cutting. I was like, fuck you. What I needed was to not be at home, to get some space to be away from everything for a while, to help myself calm down, and so I ran away. I was sleeping at a couple of houses, and while I was there the counsellor asked me to come into the school office.

According to Daphne, the counsellor did not tell her that she was bringing in the emergency mental health team to assess Daphne. “She lied to get me there and I hate her. I will not talk to her ever again cause she just like … that was wrong.” The emergency mental health team did the assessment. “They took this whole assessment of me and they told me that they wanted to get me in somewhere. And I was like, fuck you! But, of course, I sat there going, yes, yes, ok, ok.

Daphne got permission to go downstairs and have a cigarette; she had arranged for two friends to wait for her. “We bolted. I hid at my friend’s house for two hours.” The police apprehended her at a bus stop. “They put me in the drunk tank … to decide whether they were going to arrest me under the mental health act.” She tried to calm down by meditating. “I’m not going to let you see me break down. And so they drove me to the mental health thing, and they put me in this room where I can’t see everywhere and you can see in.”

Daphne disclosed that prior to this event she had attempted suicide. She had attended at her doctor’s office because she was depressed and he gave her Paxil to help with her depression. “I had an allergic reaction to it, and that is one of the reasons why I freaked out and tried to kill myself.”

Daphne was placed in “a mental hospital,” and her mother came to visit her. “She showed up with a whole bunch of stuff. She didn’t want me there.” Her mother brought her special “teddy,” cigarettes, and Cheezie’s. “She was pretty sweet about it. You know, I had people show
up at the hospital for me everyday.” She phoned her grandmother in Ontario because “she has been in mental hospitals, too.” “Dad and Mom put her in there one time.” She did not tell her father that she was there because “I didn’t feel they needed to know.” “I was really embarrassed about the fact that I had to go to this point when I didn’t feel it was right.”

Daphne did eventually tell her father. He came to see her at the hospital. “He showed up and asked me to live with him. That was really sweet, too.” Daphne decided to move in with her father although she had to spend a few days deciding if it was the best move for her.

I had to think about it, like, was this really what I wanted. And if I could live with them because Dad’s sickness is really hard for me to deal with sometimes. It is like having your mortality shoved in your face every day. It is, like, you’re going to die.

Daphne recalled that when she learned about Huntington’s Disease at fourteen she “totally denied it.”

I really didn’t think about it. If I dwelled on it I would get really sad and you know it just wasn’t really worth it at that point to actually sit down and think about it.

Daphne lived with her father for one year. She recounted how she had always struggled in her relationship with him because of feeling abandoned.

I still don’t understand how you could leave your kids. I can’t remember when he left. He didn’t come back until, like, we were eight or nine. I have tried to forgive him for this, but why should I take him back into my life when he could leave me for so long?

She also acknowledged that she has “always been a little bit scared of him.” She traces it back to a time when her father was so angry that “he ripped his shirt and the buttons off and stormed around and yelled and screamed and stuff.” “I was little and you know it scared me. And I have always been a little afraid of him.” Daphne would often take friends with her to her
father’s house because “if he hit me or something then they can do something about it . . . I was never sure how he was going to react.”

During the year she was with her father he lost his driver’s license. “Dad’s car was his freedom. If he was mad or something he would drive.” After the license was revoked he became “a total recluse, like, stuck in the house and would not leave.”

You know I never could understand how he could sit around the house all day when he knew that the end was coming. I understand that he has depression. So do I. Right? But still, I would have gotten out in the world and done something, not sat around in the house all day.

Daphne moved out at eighteen because her father separated from her stepmother. “He could not support us and so I have been living on my own for awhile. I bummed around from house to house.”

During the time Daphne lived with her father she attended a local HD Society meeting.

It helped me sort of identify with people who had been in the exact same situation as me. It was also really hard for me because you are standing around a lot of people who are dying . . . I mean, it’s not the best to look at it this way but it is sort of the truth. They are dying because they have HD.

It was at this meeting that Daphne was able to talk to people about getting tested.

It is hard to find people who are in my age group and have some kind of sense of understanding of what’s going on. I mean a lot of my friends have trouble identifying with me when I talk about stuff like this.

Daphne tries to prepare her friends before they meet her dad for the first time.

I describe it a lot like Alzheimer’s because it has . . . somewhat similar effects. It is gene oriented, like something having to do with the genes. I am 100% sure. And that it is hereditary, so like I can possibly get it, or my brother. It is basically movement-oriented. It is like the movements getting slowly sloppy and you shake around a lot. Um, short-term memory goes first. You have to remind them of things constantly. They yell at things, but he doesn’t mean to be so loud . . . it is because he is having trouble getting his thoughts in. That is basically how I describe it.
As the interview continued Daphne admitted that she was “still pretty depressed.” Daphne has decided that she is “not ever going back to the mental hospital.” Yet as the conversation continued she admitted that she “could end up back there again really easily.” She recounted the various medications she has used to deal with her depression. Her father’s doctor prescribed Effixer, and she felt that “it did work for a while but I don’t want to be drug dependent.” “I don’t want to live my life [with] the only way making me happy, is being on drugs.” She feels that “going out with Mother Nature” and meditating is helpful. “It is to find your centre for a while and sit there and get totally zoned. It is the best thing.”

Daphne feels “very close” to her brother. “The one thing that we have always had is each other through all this craziness.” She said that her brother is going to be tested. She is supportive of his decision because “he is of the mind set that he would rather know now.” She feels she would rather wait. “I don’t think I could live with knowing. I would get into a deep depression and never come out. That is not what I want to do with my life.” She confided that she has always thought that she has the gene and that her brother does not. She feels she has a good chance of having HD because “it runs in our family so strongly.”

Daphne moved back with her father at age nineteen. Her grandmother is now living with her father.

It is so much easier on me because I can sort of back off and go to my room and don’t always have to be around when I am fighting with him. Nana is happy because she has someone to take care of, and Dad is happy to have someone take care of him.

Daphne feels close to her grandmother. “It’s nice being with Nana because Nana and I always got along really well. Like we are two peas in a pod.”

When asked about the future, Daphne’s first response was “I want to have kids.” Daphne said that the geneticist in Ontario as well as “some of the other people I have talked to” suggested that she not have children.
Like the guy who was way back when, at that first meeting. He said, well, it was hereditary. I know that Nana was told to not have kids. I know that Mom and Dad were told not to have kids. Look, this runs in your family.

Her Nana told her that her parents had thought about “aborting” her. Daphne believes that they “have never regretted not aborting me.”

Daphne stated that if she decided to have children she would get tested.

It is hard because it has changed my outlook on life. Can I really be a Nana? Because she was warned not to have kids, and she has to watch her son slowly die of HD. Do I really want to put myself in that kind of a position? And is it really fair to the child? Is it fair to me? Or is it just being selfish to want kids? It is a real controversy in my head.

Daphne believes that this dilemma in her life has had a positive effect. “It makes me think about things, and I actually have to plan which is good for me because I don’t plan.”

I have had several phone conversations and an additional meeting with Daphne. She has decided that she wants predictive testing, and feels that it is unfair that she has to wait for six months before there is a space for her in the program. Her mother has phoned me with concerns over her daughter’s ability to deal with the test results. I am hoping to meet with Daphne and her mother to assist them with this issue and to help them keep the lines of communication open.
Emma: Age 17

- Moved to new community
- Dad had appointment with MD – Mom told me Dad had HD
- Mom explained HD

- Mom died of cancer around my 16th birthday
- Found out Aunt Gail had HD
- Jason broke up with me
- Got assistance with counsellor
- Found papers on family history with HD
- Visited Vancouver for 2 months to get a break

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- Mom’s cancer is back
- Notice Dad’s shaking
- Uncle died of an overdose of drugs
- Left school end of grade 9
- Made self believe Mom would live longer than Dad
- Met boyfriend Jason
- Met social worker

- Made plan with social worker to move out
- Attended HD conference in Halifax
6. Emma: (Age 17, younger brother, Dad has HD)

I arranged to meet Emma at a national HD conference through the social worker for the HD Society. Emma lived in a remote rural community, and her father had agreed to deliver her to the hotel where I was staying and where the conference was held. The social worker had arranged a room for Emma, and Emma was to attend the conference and the adolescent group I facilitate for the society.

I received a message on my phone to meet her in the lobby on the afternoon of our interview. She appeared uncomfortable with her father and sat far away from him. He told me that he drove her to meet me because the social worker had promised I would give him thirty dollars. I gave him the money and he left immediately. It was evident that Emma was tired and hungry. She had no money and only a plastic bag with a change of underwear and a box of cookies. I bought her a snack and a beverage before we went upstairs to the interview. She was frightened, and we spent some time just getting to know each other before I began the interview.

“It's not the Huntington's that bothers me; it's my dad's temper”

Emma learned about HD at fourteen when her mother told her that her father had an appointment with the doctor.

She explained to me that there was a disease that ran in the family and that my dad was going to an appointment to see if he was eligible for this disease.

Emma felt that it was “no big deal. Her father did not talk about it with her “so it was sort of a secret sort of thing.” Her mother told her that it was “a disease that affects your nerves and it makes you a little weird.” She knew that her grandmother had died from a disease, and she believes her aunt has HD. “She doesn’t talk about it but we all know she does. What I see in my Dad, I see in her.”
Emma did not see any symptoms until she was fifteen and her father started “shaking.” She spoke of her father’s temper as “an unbelievable temper for years, and years, and years.” Now she says that his temper is “worse in different ways.”

Like he used to be abusive but now he is just verbally abusive. Little things I say to him he twists around. He gets mad about them all the time. Like he is always mad. He is never happy.

Emma does not like talking about her dad and would rather talk about her mother. “She’s always been a happy person. She’s not angry all the time.”

At the end of grade nine Emma dropped out of school to help her mother who had been diagnosed with cancer.

She had a cancer tumour in her head. It didn’t start with that. It started with breast cancer. It went to her hips and then it went to her head and to everywhere.

When asked how she learned about the cancer and the course of her mother’s disease she said that no one gave her information. “I know things from what I hear. I had to go through it with my mom. I knew what she is going through because she would tell me . . . we were very close.” She felt she was alone in caring for her mother until home care became available in the last year of her mother’s life. “It was pretty much me. I was the only one that was really there.” When questioned if her father could assist she replied, “He was there then but he’s got his disease to worry about. And having her to worry about is even worse.”

Emma hoped that her mother would out live her father.

I made myself believe that she would live longer than my father. She was doing well for a while. Like, she got all her breast cancer stuff out or whatever. The doctor said that they got it – they took it out. But obviously it didn’t work

Emma’s mother died in April of 2000. Emma had just turned sixteen. “I felt like somebody just ripped out my heart. I thought I’d just die with her maybe.”
When exploring how Emma has coped with her mother’s death and her father’s HD she explained that she was strong and had to “hold everything in.”

Well, that’s what I do most of the time. Because, you know, I have to do it for my dad, my brother, and for everybody else. I’m the oldest. I don’t have disease. My dad has disease. My brother is too young. I’m right in between. I have to do practically everything. I have to be the mother.

Emma has found writing in a diary helpful. “I have a diary that I just get everything out and into it. It’s the only person – it’s the only thing I actually talk to. It makes it easier to write things down when I am not feeling good.” Emma also finds singing helpful. “When I’m not feeling good I’ll put on a CD and I’ll just sing.” Emma had a friend named Jason who she could talk to but “he wanted to be more than friends.” “When I finally went out with him it didn’t work out. So, we’re not friends no more.” It was a significant loss for Emma because he was the only one she talked with after her mother died.

Shortly after she ended her relationship with Jason a social worker with the ministry was able to get a trip funded to Coquitlam so that Emma could have a break. She had friends in Coquitlam and was invited to stay with her mother’s best friend. She loved to “just sit around in Martha’s room and pig out on food and watch TV all night.” Emma found it difficult relating to Martha’s husband. “If he tried to give me a hug, get close, it’d be like, get away from me. I’ve never had a father figure, and it’s weird thinking that some person actually loves you that’s a guy.” Emma stayed with Martha for two months before she returned home.

Emma recalls that her father has embarrassed her “forever.” “With the disease, it’s even more embarrassing. . . . I’ve got to explain to people because then it makes me feel, I guess, a little bit better that they know he’s not normally like this.” When she meets somebody new she “automatically” tells them. “I just say he’s got a disease. He can’t help the way he acts. Because he’s really embarrassing and he says the wrong things and he gives people the wrong
impression.” Sometimes Emma feels that her father is rude and it is not because of HD; she
blames it on HD “as an excuse.” She generally tries to keep her friends “away from my home.”

Emma gets support from her cousin Louise and the social worker that is affiliated with
the HD Society. Louise’s grandmother is Emma’s father’s older sister. Louise does not have HD
in her family because “her mother and her grandmother don’t have it.” Emma does not share her
grief or her anger at her father with Louise.

I want Louise just to be Louise. I don’t want her to be my little counsellor girl or
anything like that. I just want one friend who doesn’t know a lot and somewhere I
can just go and have fun and not worry about anything.

Emma has a close connection to the social worker from the HD Society. “I always call
Joan. I talk with her every couple of days.” Joan was the one who arranged home care for
Emma’s mother, and who assists her in problem-solving situations that arise at home. “Joan is
basically the only one I have to talk too.” Emma does not always agree with Joan’s conclusions.
They disagree about the root cause of Emma’s father’s abuse. “Joan tells me that it is HD, but I
don’t really think it is.” Emma has memories of abuse all her life, and recounts that the physical
abuse stopped “probably about seven or eight years ago.” “Some of the abuse might be HD, but .
. . you can’t use that as an excuse all of the time . . . there are parts of him that are abusive.”

Joan is helping Emma find another place to live. “Joan is going to help me find
somewhere to live – to get away from everything. I’ve had just about enough.” Her dad does not
know about the move.

He’ll explode. He’s going to get so angry. It’s just you got to assume the worse.
He’ll scream. He might even kill me. Well, I know he won’t kill me but I’m just
saying that is how bad it will get.

When asked how she will handle her father’s temper when he learns of her departure she
replied, “I’m not telling him.” “Joan’s going to do it.” When Emma needs courage she “looks up
at the sky” and talks to her mom.
Emma would like to have predictive testing so she can “plan my life out.”

I want to get tested now but nobody will do it. I found papers that say you can get it from seventeen and up, but then I go and I ask Joan and all them people and they say, no, don’t do it now, don’t do it now. I guess they’re afraid that if I find out I have it that it’ll be just one more thing that I have to deal with. And Joan thinks I have a little too much stress to be thinking about getting tested.

Emma has dreams of “living in Beverley Hills and getting a nice house, nice car, have a nice family, two or three kids.” She wants to become a “professional singer.” “My mom was going to get me singing lessons, but then she died. She didn’t think I had the talent, and wanted me to think that I need to practice more to be good.”

When Emma thinks about the future she does not think about Huntington’s Disease.

I think I’d be a nice person. See, it’s not the Huntington’s that bothers me, it’s just my dad’s temper that bothers me. If he were a nice person with HD it wouldn’t bother me. I wouldn’t care. And that’s what I’d be like – a nice person with Huntington’s. But he’s not. Huntington’s, that doesn’t bother me at all. I don’t care. I get it, I get it. I don’t care as long as I live a long time.

When questioned about suicidal ideation, Emma admitted to thinking about taking her life. When she feels this way “I sing or do my journal or just, you know, have cigarettes.” She feels that “it is probably my mom” that gives her the faith to keep going. “I want to live, but just not in my situation.”

After the interview I took Emma downstairs to the restaurant where she ate an entire pizza and talked about her future dreams when she moves into town. I had never eaten with anyone as hungry as Emma; I left the restaurant with a deep appreciation for the struggles this young girl deals with on her own. I arranged for her friend and cousin Louise to meet with her and had several visits with her during the conference. On the last day of the conference Emma sought me out to give me a hug goodbye and to tell me how important it was for her to tell her story.
Tessa: Age 13

- Oldest sister leaves home to go to university
- Patricia-Lynn came into our lives
- Mom stopped going out of the house
- At Christmas, Mom fell down the stairs
- Mom got live-line machine
- Oldest sister moves to London, Ontario
- Family dog died
- Class assignment on Mom
- Made secret plan to run away if Patricia-Lynn moves in

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- Mom told me about her HD
- Realized Mom was different from other moms
- Started to pull away from Dad
- Sisters had big fight with Dad about Patricia-Lynn
- All sisters asked Dad not to be so close with Patricia-Lynn
- Mom falls down stairs again
- Worry about Mom dying
Tessa and two of her sisters attended the national HD conference, and through the social worker we had arranged meeting times for the interviews. Tessa, the youngest sister, wanted the first interview. She was quite excited about participating and immediately informed me that she had never had the opportunity to talk about what life is like for her. She wanted to speak first because she felt that she was always the last to have any input in her family.

“You never really can tell when she’s sad ‘cause I’ve never seen her cry.”

Tessa started the interview by saying that when she was ten her mother told her that she had had HD since Tessa was three years old. Although she was comfortable asking her mother about HD she also said that she “never talks about it.” When asked if she talks to her sisters about HD she replied that she talks to them “once in a while.” “But we don’t really talk about it. We’re not close.” Tessa has a dog that she has had for eight years. “It sounds kind of funny, but I always talk to him. If I’m sad and stuff, I just talk to him.”

As the interview continued it became clear that when Tessa turned ten her life began to change. Tessa started to realize that her mom was “different than everyone else.” “I never really started to realize it until a couple years ago. I don’t know . . . when I like walk places with her and stuff. She didn’t have a walker back then.”

When Tessa was ten a woman named Patricia-Lynn came into her life. “She makes everything worse. She just tries to act like my mom and everything.” Tessa has tried to talk to her sisters about Patricia-Lynn but “when I tried talking to my sisters about it, they’re just like, Tessa, shut up, leave me alone.”

She talked about a time when she was twelve and she “freaked out” at Patricia-Lynn. “I got really mad at my dad and stuff. And I just started telling them that I hated her and that she
always acted like my mom and everything. So, I just got really mad at him.” She has talked to
her friends about her feelings. “They don’t like her either. Neither do my sisters. They all pretty
much hate her.” Tessa has tried to talk to her mother about Patricia-Lynn. “I tell her how much I
don’t like Patricia-Lynn because she screwed everything up.” Tessa’s mom sometimes agrees
with her “but then she just doesn’t say anything.” “It’s hard to tell with my Mom. You never
really can tell when she’s sad ‘cause I’ve never seen her cry.”

Tessa says that her friends “get to spend more time with their moms.” Tessa would like to
do more with her mom but in the last three years “she can’t really go anywhere and she doesn’t
talk a lot.” In some ways Tessa is glad that her mom can’t go out as much because “some people
are really mean and they make fun of a lot of people.” “I’m scared that they are going to make
fun of my mom because you see people walking on the street who have something wrong with
them and then they start making fun of them.” Tessa doesn’t talk about her relationship with her
mom with her friends. “I don’t really talk to them about that. I just talk to them about Patricia-
Lynn ‘cause I like tell them everything.”

Tessa began to worry about her mother dying “last year.”

Pretty much when I realized that Patricia-Lynn was there. I don’t want my mom
to die for a couple of reasons, but I don’t want her [Patricia-Lynn] to come into
my life even more than she is. I have a feeling she’d like move in with us and
stuff. I don’t want that. I’d move out if she moved in with us.

Tessa was twelve when her mother fell down the stairs and “we had to take her to the
hospital to get stitches in her forehead.” She was with her friend and her sister Margaret in her
room at the top of the stairs. “We were at the top of the stairs and I heard her fall and ran to the
stairs and got her.” She was at the bottom of the stairs bleeding; “not, like, a lot but she was
bleeding.”

Mom has a lifeline thing around her neck, and if she falls or something and is
home alone she presses the button and an ambulance comes. So, we had to press
help on it and they called us and we got an ambulance and they came and took
her.

With resignation, Tessa stated that her dad was at work. “That’s where he usually is. My
dad’s never home for supper. He gets home, like, after we eat. He works a lot.”

Tessa participates in caring for her mom.

When I come home from school, after field hockey practice, I feed her and stuff.
Or my sisters and me take turns feeding her because she can’t lift up a fork
anymore and she shakes too much.

Homemakers come to the home twice a week and “give her a bath and help her get
dressed.” “She has people that come and make her supper and stuff. On Friday my dad makes
her food but he works from 8:00 a.m. until 6:00 p.m.” Tessa says that her mother watches TV all
day. “Mom watches TV and does the laundry.” When Tessa comes home from school “we just
ask her how her day went and that’s pretty much it.” Tessa says that she is “pretty much the only
one” who doesn’t have a parent watch her field hockey games. She says her dad is “too busy and
never comes.” Once her older sister, Barbara, came, but three years ago Barbara moved away to
school so now no one comes.

Tessa remembers a time last year when Barbara “had a big fight about Patricia-Lynn.” I
was upstairs and I could hear this yelling downstairs and Patricia-Lynn was there too. That’s
another thing I don’t like. If I tell my dad something, and I tell him not to tell anyone, he’ll tell
her and stuff. She said that Kathy also had a big fight about Patricia-Lynn and moved out of the
house temporarily.

Since Tessa was ten she has gone with her mother and father to a cottage that they share
with Patricia-Lynn and her family. Last summer Tessa became uncomfortable with how the
seating arrangements were at the cottage. “Like my dad sits with Patricia-Lynn and then it’s my
mom. I told them I didn’t like it and then they said they’d change it but they never did.” Tessa says she has “lost her courage” to talk with her dad.

I’m scared to talk to him about everything. I’m scared to tell him that I don’t like her and stuff. He’ll probably get mad and he usually does. When he is really busy and stuff and I ask him something he’ll just tell me to leave him alone.

Tessa said that her next-door neighbour “always brings up Patricia-Lynn.”

She’s always like, Isn’t Patricia-Lynn your father’s girlfriend? And that makes everything worse. HD doesn’t bother me that much because it was always how I was raised. It’s not HD that’s hard for me, it’s Patricia-Lynn.

What Tessa knows about being at risk for HD is that “I have a 50:50 chance of getting this disease.” When asked how she learned about being at risk Tessa said, “I think I was reading a pamphlet one day.” Tessa doesn’t think about getting HD, although she says she “could get it soon.” “Isn’t there someone who got it who was, like, a teenager who had it?” Tessa needed information on HD. Time was spent explaining Juvenile Huntington’s and average age of onset.

Tessa would tell other young people at risk to “talk to your mom about it.” “It’s better if you talk about it but I just don’t have anyone to talk about it to. I would tell them that I find it’s easier to talk to friends about things like that.”

She recently had a class assignment where the students had to pick someone who “influenced us in the past year.” Most of her friends chose some famous people but Tessa chose her mother. “I just think she’s, like, really great and stuff for what she’s going through.” When asked if she got help on the project from her mother or anyone else in the family she replied, “I didn’t ask my family anything.” “I just did it myself. I plopped it on my dad’s desk by accident and my dad read it after I did it.”

Tessa wants to be a dentist, “but if I got the disease I wouldn’t be able to.” “So, I don’t know. Well, after a while, I’d get too shaky.” Tessa has not talked with anyone about her concerns for her future. “I haven’t talked to anyone about anything.”
Tessa and I have kept up communication on the Internet. She emails me during times of difficulty, and sees me as someone who will listen to hear concerns in confidence. The problems have not had anything to do with her mother’s HD. The problem for her is her father’s relationship with Patricia-Lynn.
Margaret: Age 15

- Birthday party and Mom falls down stairs
- Two parents of kids at party say Mom was “drunk”
- Started to cook and clean for Mom
- Patricia-Lynn came into our lives
- Learned HD was hereditary
- Mom’s speech slurred and falling down a lot
- Dad starts HD chapter in town
- Low self-esteem
- Shift in self-esteem
- Feel better about myself
- Big family fight at Christmas
- Dad reveals agreement he and Mom have about Dad and future partners

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- Told about HD
- Sent to counselling
- Homemakers start to come
- Help Mom bath and shaving legs
- Girlfriend’s Mom helps with personal issues
- Still low self-esteem
- Learned not to let peoples’ stares at Mom bother me
- Started feeding Mom
- Boyfriend discovered Dad was intimate with Patricia-Lynn
8. Margaret: (Age 15, 3 sisters, Mom has HD)

Margaret is the second youngest sister of four siblings. She arranged to be the second sister interviewed, and, like her sister, was quite excited to have the opportunity to share her story. She believes that no one knows how she feels, and that she is more isolated from her sisters because she has a good relationship with her father. She found that the interview gave her the opportunity to explore many confusing issues in her life.

“I can’t remember my mom as healthy. I’ve never had a mom like everyone else.”

The first time that Margaret realized that something was different in her family was at her seventh birthday party.

We had just moved into town. Nobody was aware of my mom’s disease. She had fallen down the stairs and two of the mothers were there and they took their daughters home because they thought she was drunk.

When asked how she knew her friends’ mothers thought her mom was drunk she replied, “One of them told me at school the next day.” “You know how little kids are. They don’t keep anything, they splat it all.” At that time Margaret “didn’t really know what was going on and why mom had fallen down the stairs, but I knew she wasn’t drunk. I knew that much.” She finds it “hard to remember” her mother without HD since she has “lived with it all my life.”

At ten Margaret remembers going to counselling because “I was upset.” “I didn’t know how to tell my friends what she had, and I was really emotional and sensitive and wasn’t sure how to deal with it.” Margaret began to “open my eyes and . . . to realize everything.” “Mom was falling more often, her speech was slurred and I could just tell. I talked with my parents a couple times about it.”

When Margaret was eleven her father started a chapter of the HD Society.
Since he is the president he’s got all these pamphlets and posters and things like that in his office. So, I often saw bulletins on the wall and posters, things like that. I remember a couple of times I was reading them and I remember crying about them sometimes.

When Margaret was in public with her mother “there’d be people just staring.”

You’d be like, stop staring! But they’re just strangers. They didn’t know what was going on but it really bothered me. But it’s only been for about a year or so that I just let it not bother me because that’s how it is.

Margaret began helping her mother when she was eleven years old by cleaning around the house and assisting her mom with her bath. She shares her duties with her sister Tessa.

Last year I noticed that mom couldn’t get the food into her mouth by herself because she was shaking so bad, so I suggested to her that I could try feeding it to her. She said, ok. She liked that and wanted us to try that. So we’ve been doing it ever since the winter. We feed her all of her meals.

When asked why she was the one who understood that her mother needed assistance with eating Margaret replied, “I’m the one who sees my mom the most.”

I’m usually the one that’s less involved in school activities. I always help her the most because my little sister Tessa has a lazy streak in her. When my mom asks her to do things, she’ll be like, urrgh, ok. We always tell her not to do that because we don’t want her to make my mom feel like she’s bothering us.

She said her dad works a lot but does the “most he can.” He buys groceries, grinds her mother’s dinner, feeds her in the morning, and buys her “lots of sweets.” Margaret feels close to her father.

My dad has always been the comforter in the family. My mom can watch anything, and she’s a very strong woman, but she doesn’t cry at all. I’ve never seen my mom cry, which is hard to believe considering she’s got HD. If I had HD I’d feel kind of a failure because I couldn’t do so many things. The reason I go to my dad is probably because he makes things a lot more clear and he’s able to express things more clearly than my mom could.
Margaret feels she is “very personal” and does not like to talk about her “home situation.” She speaks confidentially to her best friend, Tina, and her boyfriend, Bill. She does not tell her best friend as much as her boyfriend because “she tends to judge my dad.” “Tina doesn’t like Patricia-Lynn, and she doesn’t really like my father.” Bill does not state his opinion of her parents or Patricia-Lynn. “I don’t know if Bill likes Patricia-Lynn but he never says anything to me. If he thinks it, he doesn’t tell me but I always ask him to tell me what he thinks. He doesn’t judge. He just listens.” Margaret doesn’t want people to think that her family is “weird” or to have people “not like my dad or think he is bad or evil.”

Margaret says that people say that she should talk with her sisters about HD but “that’s the only thing I hate talking about.”

I’d just rather not deal with it because I have my own way of dealing with it. I think we all see it differently because of how we see Patricia-Lynn. My perspective is altogether different than my two older sisters.

Margaret feels that the problem is Patricia-Lynn not HD.

The thing is if there was nothing going on between Dad and Patricia-Lynn my sisters wouldn’t be upset. We all want my mom to be taken care of, and Patricia-Lynn is coming in and helping my mom, but it’s the fact that there is something going on and they just don’t approve of it.”

Last Christmas was a difficult time for Margaret. Barbara, Margaret’s oldest sister, came home for the holidays and “she noticed that my dad was spending more time with Patricia-Lynn than he was with my mom.” “But it’s because they’re more best friends and he just needed somebody to talk to and he was lonely. Barbara wasn’t used to seeing that and a big fight broke out.” Margaret was upstairs in her room but the voices were so loud that she heard the conversation. “I remember him saying that there were holes in his soul and she was helping him get through the tough times.” Another fight occurred between her dad and her sister Kathy.
"Kathy was too rude and mouthy to my parents and she got kicked out of the house. She came back soon and was there for Christmas but there was a lot of yelling going on."

During Easter holidays at the cabin the family shares with Patricia-Lynn, Margaret woke to a noise of a dog whining. Patricia-Lynn was not asleep on the couch she usually sleeps on; it was then that Margaret realized her father was sleeping with Patricia-Lynn.

I let the dog whine, and Patricia-Lynn came downstairs and I asked her where she was sleeping. She said she had slept in her son’s room but I know she hadn’t. My dad came downstairs and he explained everything to me. He told me that he had asked my mom that if she got really sick and he was really lonely if he could have some other kind of partner. She said she would allow him to do that while still being married. Patricia-Lynn helps my mom and she sews for her and she cooks and she cleans for her and things like that. It’s very strange and I do not tell many people.

She says that her two oldest sisters know but that Tessa doesn’t know.

You just have to find out the hard way. We find out things indirectly. It’s very hard to live with. I know I should be respectful of my parents’ decisions but I don’t feel they’re being very considerate of how their children feel about it.

Her father told her something she found reassuring: “If he could go back in time and still know that my mom was going to be sick he would still have married her.”

Margaret feels that she is “probably the only one out of all four sisters who likes Patricia-Lynn.” “I know that for a fact because I have heard them say bad things about her.” She reiterated that her father, mother, and Patricia-Lynn “work together.” “It’s not some kind of sick little triangle.” Margaret carries the bulk of the responsibilities of her mother’s care and finds it “a big relief when [Patricia-Lynn] is around because at home I’m having to cook, clean, and do a lot of things. It’s nice for somebody else to be able to do that.”

Margaret found that because her mom has HD that she didn’t “get advice on things that a daughter usually gets from her mom.”

I wasn’t seeing the typical mom and dad scene. I wasn’t seeing things that other kids my age would see. And sometimes I’d feel left out.
In grade six Margaret felt that she had “low self esteem.” Her mother didn’t drive, and her father was “often gone at meetings at night,” so Margaret would have to walk most places. People would volunteer to drive her but she felt that “people are just feeling bad for me.”

You can just sense from the way they’re looking and talking at me. They think, Oh, you poor girl, your parents. . . . I feel like they’re looking down at me, as if my parents don’t care. . . . I don’t like being judged or looked at.

Since grade seven Margaret has been able to get support from her best friend’s mother. I’m really close to my best friend’s family. Her mom is kind of like a second mom.”

She’s a very strong, confident woman, and she would always give advice to Tina and I about how to act and how to deal with things. Having that kind of influence around me had an impact on who I was, and I was just tired of being a person with low self-esteem. I was tired of trying to make other people happy.

In grade nine Margaret underwent “a big change.” “I was more confident and began to stick up for myself more at school. So, I feel much better now that I can do that because before I just let people walk all over me.”

Margaret sees that her mother’s ability to function is decreasing.

It’s something I live with, and I know that some day my mom will be gone. To tell you the truth, I will be sad because my mom is a great person, but I will also be happy because she’s really sick. I notice that day-by-day something has gotten different about her – just little things.

Margaret has never thought about a particular career but knows that she wants to do something in business.

I never thought of what happens if I do have HD. The way I see it now is that I’ll think about it when the time comes. I am not going to let something that might happen in the future affect me now.

She has seen both Barbara and Kathy go off and pursue careers, and she knows that “my parents don’t want me to stay behind and lose out on life.” “I can’t wait to move out because I’ve
done so many things on my own. I know how to do what is required around the house and one day I can be a mom and stuff like that.”

Margaret says that she would tell other teens who are coping with HD in the family that “there are going to be times you’re going to be sad, but you have to deal with it.”

You can’t let it run your life. Sometimes I have felt sorry for myself but you can’t let it interfere. I know it sounds like a cliché but its just life.

I was struck with how hard Margaret works at trying to make things all right for her father, mother, and Patricia-Lynn. When a person spoke from the podium concerning the hardships of being a risk, Margaret broke down in guttural tears and had to be held by her father through the talk. She did not look at or reach out to her mother or her sisters.

Since the conference, Margaret has used the Internet to contact me. She emails me when times are tough. She talks about Patricia-Lynn and the difficulties she has in accepting her in her life. She feels that when Patricia-Lynn comes into her life on a regular basis that it will mean that her mother will be closer to death.
Kathy: Age 19

- Told Mom had HD
- Recognized Mom behaves differently

- Moved - new community, new friends
- Homemakers
- People looking at Mom differently
- Bad time – feeling sad

- Felt like a parent to sisters

- Moved out of house to go to school

- Mom fell down stairs
- Applied for job on cruise ship

AGE

10  11  12  13  14  15  16  17  18  19

- Mom's car accident
- Mom stops driving
- Told HD is Hereditary

- Self Pity
- Two Counsellors
- Mom gets sick I get stronger
- Understanding heredity
- Started web site
- Social Worker

- Felt strong sense of self
- Patricia-Lynn enters our life
- Talked to Dad about the effects of Patricia-Lynn

- Moved back home
- Grandfather died
- Job with boss as mentor
- Break-up with boyfriend
- Fight with Dad - temporarily kicked out of house
- Learned to take one day at a time
9. Kathy: (Age 19, 3 sisters, Mom has HD)

Kathy is the second oldest daughter and the oldest daughter that attended the conference. She is a beautiful young woman with poise and an ability to talk about her feelings. She felt that she has had an easier road than her younger sisters because she knew her mother when she was healthy and was able to do activities with her. She does not believe that Huntington’s Disease is a problem for the family. Her concerns echoed those of her sisters; they were about her father’s relationship with Patricia-Lynn.

“You never think that you’d end up feeding your mother. She always fed you and then it turns around. So sometimes I’m kind of like a parent.”

Kathy’s parents told her about HD when she ten years old. “I didn’t have a clue about it before. It just went in one ear and out the other.” Her parents also talked to her about the hereditary nature of HD. “I didn’t quite understand that either at the time. Hereditary – that’s a big word for a ten year old.” She did not see her mother as having symptoms of HD, it was “just that mom was acting different.” Kathy noticed that her mother “began shaking, she stuttered and slurred and she couldn’t drive.” Kathy described “piecing it all together” as a “long process of understanding.” “I’m very visual and I just kind of watched – that’s the way I operate.” She believes that it was the same process for her sisters.

Kathy found it “sad to watch someone deteriorate over a long period of time.” She remembers her mother as “being healthy.” “Mom was a real good baker and she was a big sewer.” She realizes that her little sisters wouldn’t have any memory of her mother before the symptoms of HD. Kathy was quick to add that “it is so normal to all of us now.” “I don’t think any of us would want any of mom’s sickness taken away from us. We know that there is no cure. We just want our mom.” At seventeen Kathy was away at school for the winter and noticed a
change in her mother when she returned home. “I noticed a huge difference in that little time. It was just after that that we had to start feeding her. I think she is really starting to go downhill.”

The family moved to the East when Kathy was twelve to be close to family. The move was hard for Kathy because she left her friends. “What I found the hardest was leaving my friends, and I cried for about a year straight.” It was a time when she “started to get bitter,” and a time of “big change that I’ll remember for the rest of my life.” Kathy also remembers the move as a time when homemakers and housecleaners began to come into the home. “People started looking at my mom and I felt sad for her.” It was when her mother stopped driving that her life became affected by her mother’s symptoms because it was difficult “to get drives to places cause Dad was always working and we couldn’t get a drive to a friend’s house.”

A year after the move, Kathy began to settle into her new community and school. She didn’t tell many of her new friends about HD. “I’d just say that my mom was sick.” She told her best friends about being at risk, and she felt that they “always gave me a lot of support.” Kathy decided to start a website for teens who were at risk for HD.

I thought maybe I could help some young people because I am more aware of it. I just expressed my feelings on how I deal with it and kind of just told them mom’s story and things like that. I got a lot of responses. It was kind of self-rewarding in a way.

It was also during this time that Kathy met a social worker with the HD Society. She’s been to our house four or five times and she’ll just talk to us separately and confidentially. If we were feeling down, she’d give us a few words to boost us up, or she’d answer our questions or concerns about HD.

Kathy describes the period from thirteen to fifteen as one in which she began to “hang around a bad crowd” and became full of “self pity.”

I’d just think, why is my mom sick? Why do I have to go through this? All my friends had moms who weren’t sick, and I was always wondering why aren’t their moms sick. It would get me mad when one of my friends would get mad at their mom. I’d be like you don’t know how long you’re mother is going to be there,
and your mother is one of the best people in your whole life. She gave birth to you, how can you talk to her like that. I was pretty sensitive, and I had to mature fast.

Kathy was sent to counselling and "even tried family counseling."

It didn’t last long because I wouldn’t cooperate. I didn’t want to talk to a total stranger about my problems. So, there was no sense in going and being put on the spot.

Kathy feels that as her mother becomes “sicker” she has had to become “stronger.” She feels that it is different for her father.

I don’t think that Dad deals with it ok. He puts on an act in front of us, but I think the sicker mom gets, the closer his friendship with Patricia Lynn gets. He becomes more dependent on someone else and I become more dependent on myself.

Kathy was sixteen when Patricia-Lynn came into her life.

It’s just odd because she walks into our home and she knows mom is sick. She takes right over . . . and she tells her kids to do what they want in our house, and she totally makes herself at home. Barbara and I have told Dad that there is nothing wrong with having a friend but not to this extent. If she is not at our house then he is at her house.

Kathy describes her father’s relationship with Patricia-Lynn as something that is “confusing” and something that she is still “unable to sort through.” “People think they are having an affair.” She is upset that her father spends money on Patricia-Lynn yet expects Kathy to give him half of her income to live at home and use one of the family’s cars. “He’s snapping at me to give him money so he can spend it on her.” Kathy feels sorry for Patricia-Lynn’s children, and recounted a time at the cottage during Thanksgiving supper when “her little girl asked Dad if he was their Dad.”

It was just shock, total shock. I don’t think that I heard anything for five minutes after that. And that was the last time I went because I just don’t want to put myself in that position.”
Kathy spoke of a fight she had at Christmas with her father over money that resulted in her leaving the home for two weeks. "Patricia-Lynn was there and she started yelling at me and my sisters and I just don’t think that’s right because she is not our parent."

During this time Kathy’s boss gave her time off to go home and see her mother while her father was at work. "My mom was really angry at Dad for doing this. I noticed that the more sick mom gets the closer Patricia-Lynn and Dad’s friendship gets." Kathy feels that if her mother did not have HD she would have “stuck up” for her in the argument.

I find she doesn’t want to argue. She kind of just sits there. The next day she said that she felt my dad was out of line. I don’t put blame on her for not sticking up for me. It’s between me and him anyway.

Kathy moved back home and said that her father “reassured” Barbara and herself that “he loved Mom and for us not to worry.” “We thought, ok, and that we will leave it for now and see.”

Kathy believes she gets a great deal of support from her employer.

I strongly believe it’s her that I get some of my strength from. I can be there and tell her anything. She tells me a lot of things. I think that’s great because we have a very special bond. She tells it like it is, whether you want to hear it or not. I needed that.

Kathy also believes that she has a “bond” and a “strong connection” with her mother. She believes it is because she knew her mother when she was healthy.

It makes a difference. My two younger sisters don’t have memories of her healthy. They grew up with a sick mom. Barbara and I saw her healthy for a while, and saw more of what happened to her as she deteriorated. [Now] I see Mom and the disease. Sometimes I look at her and I see it taking over her and then sometimes I look at her and don’t even think of it. When I am feeding her I see the disease more than I see her. When she is sitting there watching TV, I see more of Mom and not the disease.

Kathy feels that she has become more like a “parent” to her mother as well as to her sisters. “You never think that you’d end up feeding your mother. She always fed you, and then it
turns around. So sometimes I’m kind of like a parent.” At fifteen Kathy felt that she started
taking on more of an adult role in the family.

Mom and Dad trusted me to baby-sit my sisters a lot, and my sisters tell me quite
a bit. We talk about the Patricia-Lynn situation, and then all that kind of stuff. So,
we’re pretty open with each other. I wouldn’t change it for the world.

Kathy knows she is at risk but it is “always kind of in the back of my mind.” “I learned to
take each day at a time because you just get yourself in more of a mess if you don’t.” She thinks
she will worry about it more when it’s time to think about getting married. “When I am in my
twenties sometime I will worry about it but right now I’m just going to leave it.”

Kathy wants to continue hairdressing, and has applied to work on a cruise ship as a
hairdresser. She wants to pay off her student loan, and may consider taking a homemaker course
after she has worked on the cruise lines. “I have so much time ahead of me.”

Kathy ended the interview with this comment: “I’m just going to say something. I think
that all this has made me more mature. I think you’ll find that in my sisters. It was a rough little
road I took there for a bit but that’s what has made me the person I am today.”