

Fear of Health Insurance Loss Among Individuals at Risk for Huntington Disease

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Genetic testing in Huntington disease, an inherited ultimately fatal neurodegenerative disorder, is infrequent despite wide availability. Factors influencing the decision to pursue testing are largely unknown. We conducted a prospective longitudinal observational study of 1,001 individuals in North America who were at risk for Huntington disease who had not pursued genetic testing prior to enrollment. We evaluated the rationale for remaining untested at baseline, determined the concerns of those who eventually pursued testing, and assessed the population's psychological attributes. We contrasted responses between those who did and did not pursue testing, and between United States and Canadian residents. The principal reasons for remaining untested were comfort with risk and uncertainty and the inability to "undo" knowledge gained. After enrollment, 83 individuals [8.3%] pursued genetic testing. Their greatest concern was losing health insurance, and 41.6% of them [vs. 6.7% of those who did not pursue testing; $P < 0.001$] reported paying out of pocket for testing or other medical services to conceal their genetic risk from their insurer/employer. Among individuals who were tested, more United States residents [46.1%] than Canadian residents [0.0%; $P = 0.02$] paid out of pocket for health services or genetic testing. Psychological attributes were similar among individuals who did and did not pursue testing. Individuals at risk for Huntington disease who pursued genetic testing feared losing medical insurance, and many paid out of pocket for medical services. Alleviating the fear of health insurance loss may help those who want to pursue genetic testing for many other conditions. [ClinicalTrials.gov number, NCT0052143].

Key words: Huntington disease; genetic testing; insurance

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INTRODUCTION

Genetic testing is increasingly available for many conditions, including breast cancer, colon cancer, and Alzheimer disease [Chai, 2007; Herper and Langreth, 2007; Lipkin and Afrasiabi, 2007; Winslow, 2007]. Testing can lead to earlier diagnoses and improved treatment but also raises questions about potential genetic discrimination by insurance companies, employers, and others [Hudson et al., 1995; Sankar et al., 2003; Eltis, 2007; Nowlan, 2007a]. Discrimination and the psychological costs of knowing one's risk factors may influence the decision to pursue genetic testing or burden those who do pursue testing. Individuals with, or at risk for, genetic conditions feel they face discrimination in insurance access [Holmes and Rahe, 1967; Alper et al., 1994; Lapham et al., 1996; Low et al., 1998; Hall and Rich, 2000; Harmon, 2008a].

Huntington disease is a fully penetrant, autosomal dominant, inherited neurodegenerative disorder generally characterized by the adult onset of a movement disorder, cognitive decline, and behavioral difficulties. The disorder commonly leads to institutional care and eventually death within 20 years of clinical onset [Walker, 2007]. Huntington disease has no cure, and only symptomatic treatments are available. The gene responsible for Huntington disease was identified in 1993 [The Huntington's Collaborative Research Group, 1993] and direct genetic testing became available soon thereafter. However, despite the availability of genetic

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testing, testing rates among individuals at risk for Huntington disease remain low, estimated at 5–20%, with rates in the United States at the lower end of this range [Meyers, 2004; Robins Walin, 2007; Walker, 2007]. Explanations for the low testing rate include the fact that there is no effective treatment, the inability to "undo" knowledge of the disease, the fear of possible emotional reactions to a positive test, and the potential loss of health insurance [Quaid and Morris, 1993; Codori et al., 1994; Decruyenaere et al., 1997]. However, small scale studies of individuals who have been tested suggest that knowing one's gene status is beneficial [Codori and Brandt, 1994].

In a large, longitudinal observational study of individuals at 50% risk for Huntington disease who had not pursued genetic testing prior to study enrollment [Huntington Study Group PHAROS Investigators, 2006], we sought to (1) evaluate the rationale for choosing not to be tested, (2) determine the concerns of those who eventually pursued testing, and (3) assess the psychological attributes of the study population.

METHODS

Study Population

We analyzed survey responses from individuals who participated in the Prospective Huntington At Risk Observational Study (PHAROS) [Huntington Study Group PHAROS Investigators, 2006]. PHAROS is a longitudinal, observational study of 1,001 individuals at risk for Huntington disease (by virtue of having a parent or sibling known to be affected by Huntington disease) who had no clinical manifestations and who had chosen not to undergo genetic testing for the HD mutation by the time of study enrollment. The study's primary purpose is to identify early clinical precursors of Huntington disease that could be applied to controlled trials aimed at postponing the onset of illness. Individuals have been followed from enrollment (beginning in 1999) forward to the present. Consenting participants were enrolled at 43 study centers (generally neurology clinics) in the United States and Canada. The study participant's country of residence—the United States or Canada—was defined based on the location of the enrolling study center.

The Huntington Study Group designed, conducted, and has complete access to data from PHAROS. Institutional review boards at all participating sites approved the research protocols and consent procedures, and study participants at each of the sites provided written, informed consent.

Survey Instruments

Participants completed surveys at baseline and at visits occurring every 9–12 months throughout the course of the study. Demographic characteristics were collected at baseline using a medical history questionnaire.

Individuals who participated in the study had not pursued genetic testing for Huntington disease prior to their baseline visit. At each study visit, individuals were asked if they had pursued genetic testing since their last visit. For those that indicated that they had, we used the first such reported indication to determine the approximate time that testing had occurred.

The rationale for not pursuing genetic testing prior to study enrollment was determined from a survey on attitudes about Huntington disease, in which participants were given a list of common reasons for choosing not to be tested, and asked to rate the importance of these reasons in their own choice not to be tested [Quaid and Morris, 1993]. Answers were scored on a 5-point Likert scale, from 0 ("Extremely Unimportant") to 4 ("Extremely Important"). For individuals who eventually pursued testing during the study's course, we used their responses from the last questionnaire they completed before they indicated that they had pursued testing. For those who did not pursue genetic testing, we used their responses from the most recently completed questionnaire.

Insurance and employment were assessed using a modified Insurance and Employment survey [Lapham et al., 1996] in which respondents were questioned about current insurance status and the effect of their Huntington disease risk on insurance decisions. This survey was introduced in 2002 and was completed beginning at the second yearly visit. We considered all survey responses—including responses after testing—when describing payment for health care services.

We conducted several psychological surveys at the baseline visit. The Beck Depression Inventory [Beck, 1987] measures depression, with scores ranging from 1 to 50, where higher values indicate more depression (scores were based on the Beck Depression Inventory I questionnaire). The Beck Hopelessness Scale [Beck et al., 1974, 1988] ranges from 0 to 20, with higher values indicating greater hopelessness. The Miller Behavioral Style Scale assesses coping preferences and divides individuals into monitors or blunters based on their self-reported preferences for information or distraction in a variety of naturalistic stress situations [Lerman et al., 1994]. A life orientation questionnaire measured one's outlook on life [Carver et al., 1994; Scheier et al., 1994] and ranged from 0 to 24 with higher values indicating greater optimism. Tolerance for ambiguity is based on anticipated responses to ambiguous situations, and varies from 0 to 20 with higher values indicating greater tolerance. We also assessed two components of spiritual well-being [Paloutizian and Ellison, 1982; Ellison, 1983; Ellison and Smith, 2007]: a religious well-being scale and an existential well-being scale, both with ranges from 0 to 60.

At baseline we also evaluated personal beliefs about Huntington disease using a survey in which individuals reported their perceptions about their own Huntington disease risk and status. Individuals reported their self-assessed probability of carrying the Huntington gene, on a scale from 0% to 100%, and their likelihood of becoming ill in the next 10 years, scored on a 5-point Likert scale ranging from very unlikely to certain.

Statistical Methods

We analyzed the rationale for remaining untested at baseline, determined the concerns of those who eventually pursued testing, and assessed the study population's psychological attributes using responses from the surveys. We contrasted survey responses between those who did and did not pursue genetic testing subsequent to enrollment in the study and between U.S. and Canadian residents. We used two-sided *t*-tests to test for differences across groups and noted any differences that were significant at $\alpha = 0.01$ to 0.05. We did not make any adjustments for multiple comparisons.

RESULTS

Study Population

The baseline characteristics of the 1,001 individuals enrolled in the PHAROS study are summarized in Table I. The average age of study participants at enrollment was 41 years, and 69% of the study participants are women. The sample is predominantly well educated (average of 15 years of education) and white (98%). Seventy-two percent have children.

In Canadian sites, 126 individuals enrolled, and 875 enrolled at U.S. sites. Canadian and U.S. residents had similar baseline characteristics, except those in the U.S. were older (42.1 years vs. 39.9 years; $P = 0.002$) and had completed more years of education (15.1 vs. 13.8; $P < 0.001$).

Eighty-three (8.3%) of the study participants chose to be tested during the course of the study. Individuals who pursed testing had completed slightly more years of education (15.5 vs. 14.9; $P = 0.04$), but otherwise had similar baseline characteristics to those that did not pursue testing. U.S. and Canadian residents pursued testing at similar rates (7.9% vs. 8.3%; $P = 0.87$).

Table II presents the baseline characteristics of the subset of the PHAROS population who completed the insurance and employment survey, which was introduced later in the study, at least once. These individuals were slightly more likely to pursue genetic testing (9.9% vs. 8.3%; $P = 0.24$), but this difference was not statistically significant. Individuals who completed the insurance survey completed slightly more years of education than the entire study population (14.9 vs. 14.5; $P = 0.02$) but were otherwise similar.

Rationale for Not Pursuing Genetic Testing

The primary reasons for not testing overall were the inability to "undo" knowledge gained, comfort with uncertainty and risk, and the absence of an effective cure or treatment for Huntington disease (Table III). Canadian residents were less likely to report fear of insurance loss as a reason for choosing not to test than U.S. residents (2.0 vs. 2.6; $P < 0.001$). The financial costs and time involved in testing were not major issues.

Concerns of Those Who Pursued Genetic Testing

Among those who eventually pursued testing, their single greatest concern (reported prior to testing) was possible loss of health insurance. This concern was significantly greater among those who pursued testing (3.1 vs. 2.5 than for those that did not pursue testing; $P = 0.006$). Individuals who pursued testing were also more concerned about how people will treat them if they know that they carry the

TABLE I. Study Population Baseline Demographics

Characteristic	Entire Sample	Individuals who did not pursue testing	Individuals who pursued testing	U.S. Study Sites	Canada Study Sites
Number of individuals	1,001	918	83	875	126
Age	41.8 [1,001]	41.7 [918]	43.0 [83]	42.1 [875]	39.9** [126]
Women (%)	68.9% [1,001]	68.6% [918]	72.3% [83]	68.7% [875]	69.9% [126]
Years of education	14.9 [1,001]	14.9 [918]	15.5* [83]	15.1 [875]	13.8** [126]
White (%)	98.1% [1,001]	98.2% [918]	96.4% [83]	97.9% [875]	99.2% [126]
Any children (%)	72.2% [724]	72.1% [679]	73.3% [45]	72.8% [627]	68.0% [97]
Number of children, among those with children	2.8 [537]	2.8 [502]	2.9 [35]	2.8 [468]	2.2 [69]
Pursued genetic testing after enrollment	8.3% [1,001]	0% [918]	100% [83]	8.3% [875]	7.9% [126]

Sample sizes are noted in square brackets.
*$P < 0.05$ for comparison with the other group [column to the left].
**$P < 0.01$ for comparison with the other group [column to the left].

TABLE II. Study Population With Insurance Survey, Baseline Demographics

Characteristic	Entire sample	Individuals who did not pursue testing	Individuals who pursued testing	U.S. Study Sites	Canada Study Sites
Number of individuals	740	667	73	643	97
Age	41.8 [740]	41.7 [667]	43.0 [73]	42.1 [643]	39.6** [97]
Women (%)	70.% [740]	69.8% [667]	72.7% [73]	69.3% [643]	71.2% [97]
Years of education	15.1 [740]	15.0 [667]	15.5* [73]	15.2 [643]	14.2** [97]
White (%)	98.2% [740]	98.5% [667]	95.8% [73]	98.1% [643]	98.9% [97]
Any children (%)	71.8% [653]	71.6% [610]	74.4% [43]	72.3% [565]	68.1% [88]
Number of children, among those with children	2.8 [482]	2.8 [448]	2.9 [34]	2.9 [420]	2.2 [62]
Pursued genetic testing after enrollment	9.9% [740]	0% [667]	100% [73]	10.1% [643]	8.2% [97]

Sample sizes are noted in square brackets.
*$P < 0.05$ for comparison with the other group [column to the left].
**$P < 0.01$ for comparison with the other group [column to the left].

Huntington disease gene (2.5 vs. 1.9; $P = 0.008$). In contrast to those who did not pursue genetic testing, individuals who pursued testing were significantly less likely to report comfort with uncertainty (2.3 vs. 2.8; $P = 0.003$) and the lack of an effective cure as a reason for choosing not to be tested (2.1 vs. 2.8; $P = 0.002$).

At risk individuals who pursued genetic testing were more likely to pay out of pocket for health care services to conceal their risk from employers and insurers. Compared to those who did not pursue testing (6.7%), 41.6% ($P < 0.001$) of individuals who pursued genetic testing paid out of pocket for either health services or genetic testing (Table IV). Individuals who pursued testing were also more likely to report having been refused insurance coverage for some treatment (prior to testing) than those who did not (6.8% vs. 2.6%; $P = 0.05$) (Table IV). Among individuals who pursued genetic testing, U.S. residents were more likely than Canadian residents to report paying out of pocket for either genetic testing or other health services (46.1% vs. 0.0%, $P = 0.02$). The most common services people reported paying for were genetic counseling and neurological screening.

Psychological Attributes

Table V summarizes the results of the psychological surveys conducted in PHAROS and the study

TABLE III. Rationale for Not Pursing Genetic Testing

Reasons for choosing not to be tested	Entire sample	Individuals who never pursued testing	Individuals who eventually pursued testing	Individuals at U.S. Study Sites	Individuals at Canadian Study Sites
I cannot "undo" the knowledge once I am tested	2.8 [700]	2.8 [660]	2.5 [40]	2.8 [609]	3.0 [91]
I prefer to live with the uncertainty of not knowing my gene status	2.8 [707]	2.8 [665]	2.2** [42]	2.7 [615]	2.8 [92]
I am comfortable with being at risk	2.8 [699]	2.8 [660]	2.3** [39]	2.7 [608]	2.9 [91]
There is no effective cure or treatment for Huntington disease at this time	2.7 [706]	2.8 [665]	2.1** [41]	2.7 [614]	2.8 [92]
I do not believe testing will give me increased control over my life	2.6 [698]	2.7 [658]	2.0** [40]	2.6 [607]	2.6 [91]
If my risk goes up, so does the risk of my children	2.6 [683]	2.6 [644]	2.6 [39]	2.6 [593]	2.4 [90]
I would rather wait until there is a promising treatment	2.5 [697]	2.5 [658]	2.0* [39]	2.5 [605]	2.5 [92]
I am concerned that I may lose my health insurance	2.5 [698]	2.5 [658]	3.1** [40]	2.6 [607]	2.0** [91]
I already have the children I planned to have	2.3 [686]	2.3 [648]	2.3 [38]	2.3 [594]	2.6* [92]
I would be unable to cope with knowing I carried the gene	2.2 [702]	2.3 [661]	2.0 [41]	2.2 [610]	2.3 [92]
I am concerned about how people will treat me if they know I have the Huntington disease gene	1.9 [700]	1.9 [661]	2.5** [39]	1.9 [609]	1.8 [91]
I do not have enough information about testing to make a decision	1.6 [704]	1.6 [661]	1.5 [43]	1.6 [612]	1.5 [92]
My partner/relatives do not think it is a good idea for me	1.6 [697]	1.6 [658]	1.6 [39]	1.6 [607]	1.4 [90]
I think I may already be affected	1.6 [700]	1.5 [660]	1.9* [40]	1.6 [609]	1.5 [91]
The financial costs of testing are too high	1.5 [699]	1.5 [659]	1.6 [40]	1.6 [608]	1.2** [91]
The testing process is too burdensome	1.1 [705]	1.1 [664]	1.0 [41]	1.1 [613]	1.1 [92]
The testing process takes a long time	1.1 [707]	1.1 [664]	1.0 [43]	1.1 [615]	1.1 [92]

Sample sizes are in square brackets. All question are framed to ask how important each factor is for an individual not having pursued testing prior to the current visit; all variables are measured on a 5-point scale [ranging from 0 to 4] from "Extremely Unimportant" to "Extremely Important".
*$P < 0.05$ for comparison with the other group [column to the left].
**$P < 0.01$ for comparison with the other group [column to the left].

TABLE IV. Health Care Expenditures and Insurance for Individuals in PHAROS

	Entire sample	Individuals who never pursued testing	Individuals who eventually pursued testing	Individuals at U.S. Study Sites	Individuals at Canadian Study Sites
Panel A: payment out of pocket					
Paid out of pocket for Huntington disease genetic testing?	5.8% [740]	2.2% [667]	38.3%** [73]	6.6% [643]	NA [97]
Ever paid out of pocket for health services?	9.1% [734]	6.3% [662]	34.7%** [72]	10.0% [639]	3.1%* [95]
Ever paid out of pocket for either health services *or* genetic testing?	10.2% [734]	6.7% [662]	41.6%** [72]	11.2% [639]	3.1%* [95]
Panel B: refusal of insurance					
Ever been refused insurance coverage for some service or treatment?	3.1% [740]	2.6% [667]	6.8%* [73]	3.2% [643]	2.0% [97]

NA, not applicable.
Sample sizes are in square brackets.
*$P < 0.05$ for comparison with the other group [column to the left].
**$P < 0.01$ for comparison with the other group [column to the left].

participants' perception of their risk for developing Huntington disease. Overall, the differences in the psychological variables between individuals who pursued testing and those that did not were small. Individuals who eventually pursued testing reported a higher degree of hopelessness (3.0 vs. 2.0; $P = 0.01$) than those who did not pursue testing; both these scores are within the normal range. Individuals who eventually did pursue testing also reported a significantly higher perceived probability of carrying the genetic mutation for Huntington disease (52.0% vs. 43.5%; $P = 0.03$).

U.S. residents had higher scores on measures of religious well-being (45.0 vs. 38.4; $P < 0.001$), slightly higher scores on existential well-being (48.5 vs. 46.6; $P = 0.04$), and lower scores on the Beck Depression Index (3.9 vs. 5.0; $P = 0.05$) than Canadian residents.

DISCUSSION

In this observational study of individuals at risk for an inherited, fatal neurological condition, individuals who pursued genetic testing feared losing their medical insurance and frequently paid for genetic testing and other health services out of pocket to conceal their risk from insurers and employers. While the cost of the medical services paid for out of pocket was frequently small (e.g., $200 for genetic testing), some medical expenses (such as pre-implantation genetic diagnosis, in which fertilized embryos of parents at risk for HD or with the HD gene are screened and only those that do not carry the gene are implanted) were quite expensive ($25,000–$30,000). This finding echoes more general results, in which researchers have found that individuals may pay out of pocket for genetic testing for a variety of conditions to conceal their risk from insurance companies [Billings et al., 1992; Hall and Rich, 2000].

Overall, the principal reasons for not pursuing genetic testing in this population were the inability to "undo" knowledge gained, comfort with risk and uncertainty, and the lack of an effective treatment for Huntington disease. Those who eventually pursued genetic testing were less likely to report comfort with

TABLE V. Psychological Attributes and Perception of Huntington Disease Risk

Survey	Entire sample	Individuals who never pursued testing	Individuals who eventually pursued testing	Individuals at U.S. Study Sites	Individuals at Canadian Study Sites
Beck depression inventory [range 1–50] BDI 0-9 (normal range), BDI 10-15 (mild depression), BDI 16-19 (moderate depression), BDI >20 (severe depression)	4.0 [817]	3.9 [767]	5.3 [50]	3.9 [710]	5.0* [107]
Beck hopelessness scale score [range 0–20]	2.1 [881]	2.0 [803]	3.0** [78]	2.1 [770]	2.0 [111]
Miller behavioral scale score [range −16 to 16]	4.2 [789]	4.3 [741]	3.3 [48]	4.2 [684]	4.3 [105]
Optimism score [range 0–24]	17.2 [749]	17.2 [703]	17.8 [46]	17.3 [648]	16.6 [101]
Tolerance for ambiguity [range 0-20]	11.0 [741]	11.0 [696]	10.9 [45]	11.1 [641]	10.6 [100]
Existential well-being [range 0-60]	48.2 [735]	48.1 [691]	50.0 [44]	48.5 [640]	46.6* [95]
Religious well-being [range 0–60]	44.1 [723]	44.0 [679]	45.7 [44]	45.0 [629]	38.4** [94]
Self-reported probability gene positive [0–100%] at enrollment	44.1 [679]	43.5 [634]	52.0* [45]	44.5 [590]	41.2 [89]
Self-reported chance will develop Huntington disease symptoms within 10 years [5-point scale from 0 to 4; higher values indicate more likely]	2.7 [713]	2.7 [669]	3.0* [44]	2.7 [613]	2.6 [100]

Sample sizes are noted in square brackets. All variables are measured at study enrollment.
*$P < 0.05$ for comparison with the other group [column to the left].
**$P < 0.01$ for comparison with the other group [column to the left].

uncertainty or lack of an effective treatment as reasons for choosing not to be tested at baseline.

Together the results suggest a two-step decision-making process for genetic testing. The first is to identify a rationale (if any) to pursue testing (e.g., reduce uncertainty about the future, guide life planning). The second is to address practical considerations of that decision (e.g., risk of insurance loss, potential social stigma of carrying the gene for a disease). Under this decision making process, the initial decision to seriously consider testing is *not* heavily influenced by fear of insurance loss. However, once the individual has decided based on life planning and other concerns to consider testing, at that point the fear of insurance loss becomes salient.

This two stage process is consistent with what we find in this study: the perceived risk of insurance loss was large among those who pursued testing, and a large proportion of those who pursued testing took active measures (e.g., paying for health care expenses out of pocket) to mitigate this risk. Our results suggest that insurance loss emerges only as a primary concern for those who decide to pursue genetic testing. For those that do not pursue genetic testing, insurance loss is less of a concern, at least during their pre-symptomatic phase.

To address the possible risk of health insurance loss among those who are at risk for genetic conditions, President Bush has recently signed into law the Genetic Information Non-Discrimination Act (GINA) on May 21, 2008 [The White House, 2008]. Some members of Congress argued that the bill was "a solution in search of a problem," but the mapping of the human genome and the increased availability of testing for genetic conditions has made the issues addressed in the law much more salient [Slaughter, 2006; Chase, 2007; United States House of Representatives, 2007]. The bill, which goes into effect 12 months (for health insurance companies) and 18 months (for employers) from the date it was enacted [Harmon, 2008b], will prohibit health insurance companies and employers from obtaining access to genetic information, requiring genetic testing, or discriminating against individuals due to genetic risk factors. Although there is debate about whether insurance companies would use genetic information even if permitted [Nowlan, 2007b], this bill will likely limit the perceived risk of such discrimination, which is the focus of this study. Our findings suggest that GINA could alleviate a significant concern of those who pursued testing for Huntington disease and reduce their financial expenditures. Currently, at risk individuals who have health insurance appear unwilling to use their health insurance to cover some health care expenses related to their underlying genetic risk out of fear that they may lose that very source of financial and health security.

In this study, U.S. residents were more concerned than Canadian residents about losing health insurance as a consequence of pursuing testing, and U.S. residents who pursued testing were much more likely to pay out of pocket for health care expenses than Canadian residents. This is consistent with the overall experience of populations in these countries: among random samples of individuals in these countries, Americans are much more likely than Canadians to report paying out of pocket for health care [Schoen et al., 2004]. The presence of universal health insurance in Canada but not in the U.S. probably drives these differences. GINA could help mitigate these differences in the U.S. However, GINA does not protect against genetic discrimination beyond health insurance and employment.

Psychological differences seem to play a limited role in the eventual decision to undergo genetic testing. However, the psychological attributes of Canadian and U.S. residents do differ. U.S. residents have significantly higher rates of religious well-being, which may reflect the generally higher level of religiosity among U.S. residents [McCleary and Barro, 2006], and may not be specific to those at risk for Huntington disease. U.S. residents also have lower rates of depression, although this does not seem to be reflective of the overall population in these countries, or of people with other serious illnesses [Lopes et al., 2004; Vasiliadis et al., 2007].

The findings here echo, to some extent, existing literature on genetic testing. Disclosures of HD risk within families appears to be limited, in part, by the fear that others will not be able to cope with knowing of their risk [Klitzman et al., 2007]. In the early years of cystic fibrosis genetic testing, research found some insurance concerns driving limited testing, as well as similar issues with not being comfortable knowing one's risk [Fanos and Johnson, 1995]. The data here, from a very large prospective study in two countries, reinforce the concern about insurance loss and highlight the tangible actions (paying for health care expenses out of pocket) that asymptomatic U.S. residents at risk for Huntington disease take to address this fear.

Although HD is, in some ways, an unusual genetic disorder in that a positive gene test indicates certainty of disease onset, it is not unique, and the findings here may be informative about testing for other genetic disorders. Despite this, extending our results to other genetic disorders should be done with caution, taking into account possible differences across conditions.

There are limitations to the current study. First, we undertake multiple statistical comparisons here and do not adjust our measure of significance levels to take this into account. However, in many cases (e.g., comparing payment out of pocket among those who pursue testing to those who did not) the results were significant at much higher than

the 0.01 level. In addition, the results, especially those pertaining to health insurance and health expenditures, are internally consistent with one another.

Second, when comparing U.S. and Canadian residents, we are comparing individuals who are participating at sites in each country, even though they may not live in that country. In practice, most individuals in the study participated at centers near their residence. The study population also may not be representative of the population at risk for Huntington disease as the individuals recruited in this study were predominantly women [Huntington Study Group PHAROS Investigators, 2006].

Third, we do not currently have clinical or testing outcomes for study participants, which would be helpful in learning, for example, whether their self-perceptions of genetic risk are accurate. Eventually information on gene status will be available from the ongoing PHAROS study, and further analyses of the data will allow us to observe differences across individuals related to their true gene status.

Fourth, in ranking reasons for not pursuing testing, respondents were generally forced to choose one of the reasons listed. There was a free response "other" option given, but only 7% of individuals reported anything in this category.

Fifth, very generally, this is a volunteer study, and as has been noted elsewhere [Huntington Study Group PHAROS Investigators, 2006] this may lead to differential selection of participants than a random sample study. In particular, this cohort is skewed towards women, highly educated people and those who are gainfully employed. This selection may limit the degree to which we can generalize the results.

Finally, the insurance survey was introduced part way through the study, at which point there had been some attrition in the sample. The baseline characteristics of the two populations, however, were generally similar.

In sum, this study finds that fear of insurance loss is a dominant concern among individuals at risk for a genetic disorder and that individuals will act to conceal this risk from their health insurers, even at considerable personal expense. As genetic testing becomes more widely available for increasingly common medical conditions [Herper and Langreth, 2007; Topol et al., 2007], these issues are likely to take on heightened importance. Federal legislation (GINA) may alleviate some of the fear and burden that many individuals at risk for genetic conditions currently bear.

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