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## Health Screening Behaviors among Adults with Hereditary Hemorrhagic Telangiectasia in North America

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### Abstract

**Purpose**—This study aimed to identify factors that influence screening behaviors of adults with hereditary hemorrhagic telangiectasia (HHT).

**Methods**—Participants with a self-reported diagnosis of HHT were recruited from the HHT Foundation International, Inc., the “HHT Awareness” Facebook group, and six HHT clinics. A cross-sectional mixed methods survey was administered to investigate the relationships among the Health Belief model constructs, the domains of illness representations, and HHT-specific screening behaviors consistent with recommended guidelines.

**Results**—A total of 320 participants reported rates of cerebral arteriovenous malformation (AVM) screenings, pulmonary AVM screenings, and HHT annual checkups that were 82.0, 67.1, and 56.5%, respectively. Logistical regression analysis showed perceived barriers ( $\beta = -0.114$ ,  $p < 0.001$ ), perceived susceptibility ( $\beta = 0.117$ ,  $p < 0.05$ ), treatment control ( $\beta = 0.078$ ,  $p < 0.05$ ), and emotional representations ( $\beta = 0.067$ ,  $p < 0.05$ ) were significant predictors of HHT screening. Open-ended responses revealed perceived barriers to screening, including a lack of healthcare providers (HCPs) familiar with and/or knowledgeable about HHT.

**Conclusion**—Our results reveal sub-optimal screening rates among adults with HHT, and identify several factors influencing these behaviors. We suggest that there is a need for increased

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provider education regarding HHT as well as approaches that providers can use to improve screening adherence.

### Keywords

hereditary hemorrhagic telangiectasia (HHT); screening behaviors; perceived barriers; Health Belief Model (HBM); Illness Representation

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## Introduction

### Background

Hereditary hemorrhagic telangiectasia (HHT) is an autosomal dominant multisystem vascular dysplasia characterized by arteriovenous malformations (AVMs).<sup>1</sup> The most common clinical manifestations are spontaneous and recurrent nosebleeds and telangiectases (small AVMs) of the hands, face, mouth, and gastrointestinal (GI) mucosa. However, large AVMs can occur in the lungs (PAVMs) and the brain (CAVMs), leading to stroke, brain abscess, and seizures, and in other organs, including the liver. While nosebleeds are most often the initial symptom, AVMs can occur before the appearance of mucosal/cutaneous telangiectases and leave seemingly unaffected family members at risk for life-threatening complications. HHT is a chronic condition, but with early diagnosis followed by adequate screening and treatment, often the major complications of this disorder can be avoided, and disability or even death can be prevented.

The international HHT medical management guidelines<sup>2</sup> were published in 2011, and have not yet been updated. Screening protocols in the clinical setting may therefore differ as guidelines begin to lag behind the state of the science. The 2011 guidelines<sup>2</sup> recommend screening for CAVMs in children and adults once at the time of initial diagnosis and additional screening only if there are positive MRI findings. For PAVM screening, the experts recommend Contrast Transthoracic Echocardiography (TTCE) in children and adults at time of initial evaluation and additional screening every 5-10 years if TTCE results are negative. The expert panel also recommends that patients consult with gastroenterologists to check for gastrointestinal bleeding and liver and vascular malformations and with otorhinolaryngologists (ENTs) for epistaxis assessment. Additionally, seeing a physician knowledgeable about HHT is key in the care of patients with HHT, because inadequate knowledge can raise barriers to prevention, diagnosis, and treatment. Research to date suggests that nonadherence to screening or preventive treatment is an overarching problem in many medical conditions<sup>3-12</sup> and chronic genetic diseases, such as HHT, are no exception.<sup>13, 14</sup>

### Theoretical Framework

The Health Belief Model (HBM) guided the framework of this study and has been widely used to explain individual differences in preventive health behavior.<sup>11, 15-18</sup> The model is based on the principle that people will take action to prevent, to screen for, or to control ill-health conditions if 1) they regard themselves as susceptible to the condition/threat, 2) they believe the condition/threat would have potentially serious consequences, 3) they believe that a course of action that is available to them would be beneficial in reducing either their

susceptibility to or the severity of the condition/threat, and 4) they believe that the anticipated barriers to taking the action are outweighed by the benefits.<sup>15</sup>

Although Verkerk et al. assessed screening adherence for PAVMs in patients with HHT<sup>19</sup> and found sub-optimal levels, with only 57.7% of patients having undergone PAVM screening, they did not assess factors that influenced adherence. The goal of this study was to quantify and explore factors that influence screening behaviors within a population of adults with HHT, in order to identify targets for clinical intervention.

## Methods

### Study Population

Participants were recruited from the HHT Foundation International, Inc., the [www.facebook.com](http://www.facebook.com) HHT Awareness group, and six HHT Clinics across the United States that agreed to assist with recruitment, including those at Yale School of Medicine, University of Utah Medical Center, Oregon Health & Science University, Washington University School of Medicine, University of Pennsylvania, and Johns Hopkins Hospital. A snowball recruitment technique was used to recruit additional participants. Eligibility criteria were age ≥ 18 years and self-reported diagnosis of HHT. Participants who completed the survey were given a \$10 gift card. The protocol was approved by the IRB of the National Human Genome Research Institute, and all participants provided informed consent.

### Study Design

The cross-sectional survey was based on a mixed-methods approach. The quantitative component included assessment of illness representations, perceived susceptibility, perceived benefits and perceived barriers, and self-efficacy. Rosenstock et al.<sup>20</sup> proposed that self-efficacy, or an individual's confidence in one's ability to take action, be added to the HBM as an independent variable based on evidence of its independent effect on behavior change. Illness representations were evaluated to take into account the individual's cognitive and emotional perceptions of their condition. We also assessed demographic information, including gender, age, marital status, income, education, race/ethnicity, current insurance status, country of residence, parental status, and HHT-diagnostic status of children, as well as two measures constructed for this study: 1) response efficacy and 2) an assessment of HHT screening adherence. The qualitative component to the survey included six open-ended questions to further explore factors related to adherence.

### Measures

**Illness Representations**—Illness representations were captured using the Illness Perception Questionnaire (IPQ-R)<sup>21</sup>, a theoretically derived measure which asks participants to consider their own personal views of how they perceive their current illness. The items are categorized by seven domains of illness representations: timeline (acute/chronic, cyclical), consequences, personal and treatment control, coherence, emotional representations, identity, and cause, which have been validated by Moss-Morris et al<sup>21</sup> using eight different illness groups. Psychometric analysis revealed that all of the IPQ-R subscales demonstrate

good internal reliability. The Cronbach's alphas for each of the subscales ranged from 0.79 to 0.89.

**Health Belief**—The Champion Health Belief Model Scale (CHBMS) was developed with the intention of measuring the concepts of the HBM in numerous study populations and across various preventive health behaviors. The CHBMS has been shown to be reliable and valid, and there is evidence supporting content and construct validity.<sup>22</sup> The Cronbach's alpha was 0.83 for Perceived Susceptibility<sup>23</sup>, 0.85 for Perceived Barriers<sup>23</sup>, 0.65 for Perceived Benefits<sup>23</sup>, and 0.87 for Self-Efficacy<sup>24</sup>. The CHBMS was re-designed to be HHT-screening specific. The Response Efficacy scale was adapted and validated from previous work in breast cancer screening behavior<sup>25</sup> and exercise behavior to reduce coronary heart disease.<sup>26</sup> In this study, a four question measure assessing response efficacy focused on benefits and barriers of screening the brain and lungs and seeing a HCP knowledgeable about HHT.

**HHT Screening Adherence Measure**—The HHT screening adherence measure consisted of three items that assessed past participation in screening related to HHT, including a cranial MRI (“When was the last MRI of your brain?”), PAVM screening (“When was the last time you had your lungs screened for your HHT?”), and annual follow-up with a HCP knowledgeable about HHT. The past behavior scores for each of the screening behaviors were summed and treated as a categorical variable (High, Medium, and Low). “High” adherers are participants who fulfilled all three recommended guidelines, “Medium” adherers are participants who are late on PAVM screening or late on annual check-up but have done it in the past, and “Low” adherers are participants who have neglected at least one aspect completely.

**Open Ended Questions**—Six questions were posed to elicit additional possible barriers and benefits to HHT screening. The questions were: What do you consider to be the most important benefit(s) to having an MRI of your brain? What things could/do keep you from getting an MRI of your brain? What do you consider to be the most important benefit(s) to having your lungs screened? What things could/do keep you from getting your lungs screened? What do you consider to be the most important benefit(s) of seeing a healthcare provider who is knowledgeable about HHT? What things could/do keep you from seeing a healthcare provider who is knowledgeable about HHT?

## Data Analysis

Data were analyzed using SPSS 16.0 and the primary outcome variable was the three-category measure of screening adherence. Polytomous Universal Model (PLUM)<sup>27</sup> under a cumulative odds model was used to determine the association of illness representations and health belief model constructs with level of HHT screening adherence after controlling for potential confounding variables. Each potential confounder (all demographic variables, age at diagnosis, parental status, and HHT-diagnostic status of children) was tested as a predictor of adherence using ANOVA or PLUM, as appropriate. Relationships that resulted in a p-value  $\leq 0.20$  were included in all subsequent multivariate regression models. Backwards elimination was used to test for the association of one covariate on the outcome measure

while controlling for other covariates. Our power analysis for a logistic regression (two-tailed test) using an alpha of 0.05 and 80% power suggested that a sample size of 268 would be sufficient to detect a small effect (odds ratio = 1.5).

Brief thematic analysis was used to explore data collected from the open-ended questions. Responses were analyzed for overarching themes and then grouped accordingly. Major themes were further analyzed for subcategories and frequencies of common themes were calculated. Responses representing more than one theme were coded for each theme characterized.

## Results

### Study Participants

Three hundred and twenty surveys were received, including 311 electronic and nine paper versions. Thirty surveys were submitted incomplete, and these were still included in analyses for which survey data were available in order to maximize the amount of data in each analysis. The most cited source of recruitment for participants was through the HHT Foundation International, Inc. (82.1%).

The average age was 49.5 years ( $\pm 13.7$ ) and ranged from 18-82 years. The participants varied widely in how long they have known of their HHT diagnosis, from under a year to 69 years with a mean of 29.25 years ( $\pm 14.6$ ). The study population was largely female (72.9%), white (94.2%), not of Hispanic origin (97.2%), and married (77.4%). The majority of participants (57.8%) had obtained a college or post-graduate degree. Additionally, a large majority of participants were parents (76.7%), and of those, 12.8% reported that at least one of their children has been diagnosed with HHT. Table 1 summarizes the characteristics of the study sample.

### Illness Representations and Health Belief Model Variables

The descriptive statistics and Cronbach's alpha values for all the key predictor variables are shown in Table 2. The internal reliability of each of the IPQ-R scales was acceptable with the exception of the acute timeline and cyclical timeline scales ( $\alpha=0.61$  and  $0.66$  respectively). The percentage of participants scoring above the mid-point for each scale of the IPQ-R and HBM scale is also shown; this provides an indication of the proportion of participants holding particularly strong views about the construct being measured by each particular scale. For example, most participants reported a high degree of personal understanding about their condition, with 77.3% scoring greater than the mid-point on the IPQ coherence scale. Almost all participants scored greater than the mid-point on the IPQ acute timeline scale, indicating that they feel their illness to be chronic, which is true of HHT. The majority of participants also perceived their HHT as having particularly severe consequences on their lives. Single-item analysis revealed that 84% endorsed that "My HHT is a serious condition," and 67% of participants agreed that HHT has major consequences on their life.

Exactly 50% of participants scored above the mid-point on the *emotional representations* scale (assessing negative emotions associated with HHT), with reported feelings including

depression or upset (35%), anger (29%), fear (41%), and anxiety (48%) regarding their condition.

### Reported Adherence to HHT Screening

When asked about past cranial MRI behavior, 82% reported having had a cranial MRI at some point in their lifetime, 17.3% reported having never had a cranial MRI, and 0.7% were unsure. When asked about past lung screening behavior, 67.1% reported having had screening for pulmonary PAVMs within the last 5 years, 18.1% had screening 5-10+ years ago, and 14.9% have never had their lungs screened or were unsure. The final component of the screening adherence measure asked about past follow-up with a HCP familiar with HHT. The majority of study participants (56.5%) have seen such a HCP within the last year and 9.1% have never seen a HCP knowledgeable in HHT. Ultimately, 41.3% of the study population reported having fulfilled all three of the recommended guidelines, and were subsequently categorized as “high adherers” and 35.3% reported having neglected at least one guideline completely, and were categorized as “low adherers.”

### Predictors of HHT Screening Adherence

Multivariate regression analysis revealed that positive relationships were observed between screening adherence and the variables of *susceptibility*, *emotional representations*, and *treatment control*, such that as each increased, the probability of being in a higher category of screening adherence increased. Conversely, a negative relationship was observed between screening adherence and *perceived barriers* to screening, such that as *perceived barriers* increased, the probability of being in a higher screening adherence category decreased.

As shown in Table 3, of the potential confounders, level of education was the only one found to be a significant predictor of screening adherence. Participants with a High School/GED or a college degree had statistically significantly higher odds of being in a lower adherence category than participants with a post-graduate level of education.

The overall model was significant ( $X^2 = 60.821$ ;  $df=12$ ;  $p = 0.000$ ) as compared to a model with no predictors. The final model exhibited a moderate fit with a Cox and Snell pseudo- $R^2$  of 0.212.

Based on the output of the final model (displayed in Supplemental Table 1) the predicted probability of being in different levels of adherence based on each of the predictor variables of susceptibility, barriers, treatment control, and emotional representations, while controlling for all other predictor variables (held constant at respective means; thus, the middle column of the table is identical for each variable at its mean) and the confounding variables of education, income, and insurance status.

### Perceived Barriers to HHT Screening

Two hundred fifty-four participants (79.4%) completed the open-ended questions regarding barriers to pursuing care from a HCP knowledgeable about HHT, and 267 participants (83.4%) provided perceived benefits (quantitatively summarized in Supplemental Table 2).

The majority of participants who cited no barriers/nothing often cited factors that used to be barriers or that could be, and then stated how they were able to overcome them or were in a “fortunate” situation. The next most cited barrier to a knowledgeable HCP was the apparent lack of HCPs perceived as knowledgeable regarding HHT. Participants also frequently mentioned the inconvenience of travel to an HHT-knowledgeable HCP, the time required and scheduling difficulties, and the financial burden, as this participant explained:

*So few doctors are knowledgeable about HHT that it is nearly impossible to find one who knows much about the disease. The nearest center of excellence to my home is about 300 miles... It requires a day to travel there, a day or two of appointments, and a day to travel home. I don't know many people, especially people with medical bills, who can afford to take 4 days off simply to go to the doctor about a nose bleed. (21 year old female, Low)*

Two hundred forty-three participants (75.9%) completed the open-ended questions regarding factors they consider to be barriers to having their lungs screened for pulmonary AVMs, and 268 participants reported perceived benefits (83.8%). The majority (53.9%) of participants cited that there are no factors that keep them from pursuing lung screening. As one participant reported,

*“Nothing. If I had to walk to an HHT center I would still make the trip because I can't take that risk.” (21 year old female, High)*

Two hundred fifty-six (80.0%) completed the open-ended questions regarding factors that they consider to be barriers to having an MRI of the brain to screen for cerebral AVMs, and 265 participants (82.8%) provided perceived benefits. Only three factors were reported as barriers to pursuing an MRI of the brain that were unique or not as commonly cited when considering lung screening. First was the inability for some participants to undergo an MRI, with eight participants stating that they have metal devices or implants contraindicating the procedure. Second was an increased fear of the procedure itself, primarily due to claustrophobia. Third was a sense of denial as reported by this participant:

*Getting the screening is admitting that I have a life altering problem possibly. (56 year old male, Low)*

## Discussion

This study is the first to explore and quantify factors that influence screening behaviors among adults with HHT. Specifically, this study examined the roles of illness representations and HBM constructs in reported adherence to HHT screening. The rates for CAVM screening, PAVM screening, and HHT annual check-up in this study population were 82.0%, 67.1%, and 56.5%, respectively. Additionally, 41.3% of participants reported having fulfilled all three of the recommended guidelines (and were subsequently categorized as “high adherers”) while 35.3% reported having neglected at least one guideline completely (and were categorized as “low adherers”). These results illustrate not only that the overarching problem of nonadherence in chronic medical conditions applies to the HHT population, but also that the extent of adherence varies across specific recommendations.

In general, participants viewed their HHT to have serious consequences, and the majority agreed HHT is a serious condition. Participants had a heightened perception of threat and held strong beliefs regarding the chances of serious complications. However, the participants also reported relatively high levels of self-efficacy and response efficacy, demonstrating confidence in their ability to comply with the recommended screening and that the screening will reduce perceived threat. This study sample was somewhat split in negative feelings that were triggered by their condition, while feelings of understanding (coherence) regarding HHT were quite high overall. These results provide a glimpse into the previously unexplored perceptions of adults with HHT, and demonstrate that these individuals are equipped to overcome the barriers given appropriate resources.

Two unique environmental barriers were the apparent perceived lack of HCPs familiar with and/or knowledgeable about HHT and a perceived lack of effort by HCPs to learn about HHT. Inadequate knowledge about genetic conditions among HCPs who do not specialize in genetics has been previously reported.<sup>28-31</sup> Also, Bernhardt et al.<sup>32</sup> observed a similar concern among patients with HHT: many felt that primary care providers might not be adequately prepared to order and accurately interpret genetic testing for HHT. HHT is thought to be considerably underdiagnosed<sup>33, 34</sup>, and it has long been suggested that wider physician awareness of the condition and its pathology could help increase diagnosis and avoid the risks associated with mismanagement.<sup>35</sup> Another commonly cited environmental barrier was the existence of only 18 multidisciplinary centers in the United States providing comprehensive care for individuals with HHT,<sup>36</sup> making it difficult to seek care because of geographic and financial limitations.

Similar to experiences with other conditions<sup>2, 4-7</sup> emotional and cognitive barriers to screening were identified: fear and anxiety that screening may actually find something, distrust in HCPs, lack of information regarding where to go for screening, and perceived discomfort, pain, or fear associated with the screening procedure. One troubling finding was that being seemingly asymptomatic acts as a barrier in the decision to pursue HHT screening. This is a challenging barrier because many of the major complications of HHT (e.g. PAVMs and CAVMs), can remain unrecognized until a possibly life-threatening event occurs, and the purpose of screening is to intervene, where appropriate, before any hidden complications manifest.

The most commonly reported benefit was that screening will detect complications early on.<sup>8</sup> Additional perceived benefits to HHT screening included early treatment, prevention of complications, monitoring of already existing AVMs, having more knowledge about the status of one's condition, and the belief that screening can save one's life. Participants reported multiple benefits to having a HCP knowledgeable about HHT, including enhanced patient-physician relationship, communication, and level of trust.

Both HBM and Illness Representation constructs have been shown to be strong predictors of preventive health behaviors,<sup>9, 13, 18-21, 37, 38</sup> and the present study has provided support for this in an effort to explain preventive screening behaviors in adults with HHT. Of the ten independent variables included in the multivariate PLUM analyses (six illness representation constructs, and four HBM constructs), four were found to be independent significant



predictors of screening adherence: perceived barriers to screening, perceived susceptibility to the threat of health problems associated with HHT, perceived control over one's treatment, and emotional representations triggered by the condition.

The HBM construct of perceived barriers emerged as the most powerful predictor of HHT screening. When holding all other variables in the model constant at their respective means, having a maximum perception of perceived barriers can increase one's odds of being in the Low Adherence group 10-fold. Perceived susceptibility was also found to be an independent predictor of screening adherence, with those participants perceiving a sense of vulnerability to the potential complications of HHT more likely to participate in screening. These findings are consistent with the overall evaluation of the HBM.

The Illness Representation constructs of treatment control and emotional representations were also found to be positively related, independent predictors of HHT screening adherence. Regarding the former, participants who perceived very little control over their HHT treatment were less likely to adhere to screening recommendations. Regarding the latter, participants reporting more negative emotions triggered by their HHT (e.g. worry and anxiety), were roughly three times more likely to be in the High Adherence category. While this may seem counterintuitive, greater negative emotions regarding HHT may be acting as a motivating force, similar to the increasing perceptions of susceptibility and severity in the HBM. Moreover, strong beliefs that HHT is amenable to control via specific treatments may reinforce or enhance one's motivation to adhere to the recommended treatment. Overall, these findings suggest that both cognitive and emotional factors account for HHT screening behavior in this population.

### Limitations

Despite attempts at recruiting from several different sources, the majority of participants were recruited through the HHT Foundation International, Inc., a non-profit organization whose purpose is to support and educate patients, families, and medical professionals. Considering that the majority of participants were most likely members of the HHT Foundation, it is quite possible that the sample represents a group of individuals relatively knowledgeable about HHT. In addition, the study population was largely female and Caucasian, which does not proportionately represent the population of individuals with HHT. Finally, participants were asked to self-report their screening behaviors, which introduces the potential for recall bias; it is possible that self-report measures overestimate adherence, especially when compared with more direct measures.<sup>39</sup> Lastly, many of the HBM constructs and IPQ-R variables are significantly correlated with each other (see Supplemental Table 3). We acknowledge that the possibility of multi-collinearity may increase the standard errors of the coefficients, potentially obscuring an effect.

### Clinical Implications

The results of this study reveal several important concepts for clinical practice within the HHT population. First, it is important for clinicians caring for patients with HHT to know that screening rates are not ideal, and that confirming the diagnosis and making screening recommendations do not necessarily result in adherence. The screening behaviors within this

population are complex in nature, with many factors contributing to the decision to pursue screening. Second, approximately one-third of participants reported being unable to find a HCP familiar with HHT, which is especially disheartening given that the vast majority of participants were recruited through one of the primary patient resources available for individuals with HHT. Roughly 9% of participants who provided a response regarding perceived benefits of seeing a provider knowledgeable about HHT reported assuming the role of “teacher” within their patient-physician relationship, including initiating discussions regarding their HHT treatments and necessary screening.

Several approaches can be considered to improve screening adherence by attempting to reduce barriers and by considering perceived susceptibility. Our findings demonstrate the need for further physician education, which can be provided through organizations specializing in HHT and/or genetics, such as the HHT Foundation International, or the American College of Medical Genetics and Genomics. As suggested by Latino et al.,<sup>33</sup> ENT physicians may prove an excellent physician group to initially target with educational programs to improve diagnostic rates of HHT.

In addition, it may prove beneficial to assess the patient's understanding of the potential complications of AVMs. If the patient is not fully aware of the possible consequences, such information should be presented in balance with encouragement and an optimistic perspective of the patient's ability to influence such outcomes of HHT by adhering to the recommended screening guidelines. Recent research shows that the life expectancy of HHT patients may differ based on the molecular basis of the disease.<sup>40</sup> It would be prudent for future HHT research and medical providers advising patients on HHT screening to consider the underlying genetic defect.

## Supplementary Material

Refer to Web version on PubMed Central for supplementary material.

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**Table 1**  
**Description of Study Sample**

Sociodemographic Variable		Total N	Percentage
<b>Gender</b>	Female	215	72.9
	Male	80	27.1
<b>Marital Status</b>	Married	226	77.4
	Not married	66	22.6
<b>Highest Level of Education</b>	High School/GED	57	19.5
	Some College	66	22.6
	College Degree	102	34.9
	Post-Graduate	67	22.9
<b>Race</b>	Caucasian	277	94.2
	Not Caucasian	17	5.8
<b>Ethnicity</b>	Hispanic	8	2.8
	Not Hispanic	273	97.2
<b>Annual Household Income</b>	<\$25,000	25	9.3
	\$25,000-49,999	45	16.8
	\$50,000-74,999	69	25.7
	\$75,000-99,999	36	13.4
	>\$100,000	93	34.7
<b>Current Insurance Status</b>	Insurance	267	91.4
	No Insurance	25	8.6
<b>Country of Residence</b>	U.S.	231	78.6
	Outside U.S.	63	21.4
<b>Parental Status</b>	Yes	227	76.7
	No	69	23.3
<b>Child's HHT Status</b>	Yes	29	12.8
	No/Unknown	198	87.2
<b>Demographic Variable</b>		<b>Mean (SD)</b>	<b>Range</b>
<b>Age (years)</b>		49.5 ( $\pm$ 13.7)	18-82
<b>Years since HHT Diagnosis</b>		29.25 ( $\pm$ 14.6)	0-69
<b>Recruitment Source *</b>		<b>Total N (%)</b>	
<b>HHT Foundation direct mailing</b>		100 (31.7)	
<b>HHT Foundation email notification (E-blast)</b>		82 (26.0)	

<b>Sociodemographic Variable</b>	<b>Total N</b>	<b>Percentage</b>
<b>HHT Foundation website</b>	77 (24.4)	
<b>Family Member</b>	33 (10.5)	
<b>Facebook “HHT Awareness” Group</b>	17 (5.4)	
<b>HHT Clinic</b>	4 (1.3)	
<b>Friend</b>	2 (0.6)	

\* Note: N=315; five participants did not report source of recruitment

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Table 2

## Scale Descriptives

Variable	Number of items in scale	Cronbach's alpha	Mean	Possible Range, (Observed)	SD	% scoring above scale mid-point
<b>Illness Representation Constructs</b>						
Acute Timeline	6	0.61	27.42	6-30, (17-30)	2.653	99.0
Cyclical Timeline	4	0.66	12.88	4-20, (4-20)	3.145	57.0
Consequences	6	0.74	21.21	6-30, (8-30)	4.507	71.7
Control	6	0.81	18.61	6-30, (6-30)	4.753	54.1
Treatment Control	5	0.76	14.49	5-25, (5-25)	3.795	43.1
Coherence	5	0.87	18.45	5-25, (5-25)	4.314	77.3
Emotional Representations	6	0.89	18.64	6-30, (6-30)	5.436	50.0
Identity	15	0.82	4.82	0-14, (0-14)	3.142	N/A
<b>Health Belief Model Constructs</b>						
Susceptibility	5	0.83	20.16	5-25, (10-25)	3.569	89.6
Barriers	9	0.71	19.79	9-45, (9-38)	5.387	8.4
Benefits	4	0.65	15.33	4-20, (4-20)	2.698	88.2
Self-Efficacy	10	0.90	40.03	10-50, (13-50)	7.201	91.2
Response Efficacy	4	0.87	16.78	4-20, (4-20)	3.018	92.5

**Table 3**  
**Multiple Ordinal Logistic Regressions: Prediction of HHT Screening Adherence**

	Unadjusted Model $\beta$ Est.	Final Model $\beta$ Est.
<b>Susceptibility</b>	0.074	<b>0.117*</b>
<b>Barriers</b>	<b>-0.096*</b>	<b>-0.114**</b>
<b>Self-Efficacy</b>	0.030	-
<b>Response Efficacy</b>	0.025	-
<b>Consequences</b>	0.011	-
<b>Emotional Representations</b>	0.050	<b>0.067*</b>
<b>Identity</b>	0.083	-
<b>Treatment Control</b>	0.063	<b>0.078*</b>
<b>Education</b>	<b>H.S./GED</b>	<b>-1.142*</b>
	<b>Some College</b>	-0.605
	<b>College Degree</b>	<b>-0.751*</b>
	<b>Post-Graduate (ref.)</b>	0
<b>Insurance Status</b>	<b>No</b>	-0.937
	<b>Yes (ref.)</b>	0
<b>Cox and Snell Pseudo-R<sup>2</sup></b>	0.227	0.212
<b>Test of Parallel Lines Sig.</b>	0.356	0.004

\*  
p<0.05,

\*\*  
p<0.001