Where Does Hope Fit In?
The Relationship Between Hope, Uncertainty, and Coping Efficacy in Mothers of
Children with Duchenne/Becker Muscular Dystrophy

By

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ABSTRACT

Background: Duchenne and Becker Muscular Dystrophy (DBMD) is a complex, progressive, and ultimately terminal condition laden with caregiver uncertainty often related to prognosis, medical management, social, and existential concerns. This uncertainty can make adaptation more difficult for mothers, yet some view uncertainty as allowing for the opportunity of positive outcomes. Literature suggests that the concept of hope may influence this appraisal of uncertainty. It is not yet fully understood how mothers of children with DBMD appraise, cope with, and ultimately adapt to their child's DBMD in light of this uncertainty or the role hope plays in the process.

Objective: To examine the relationships between maternal uncertainty, hope, and coping efficacy among mothers of children with DBMD.

Methods: Mothers of children with DBMD were recruited through the Duchenne Connect Registry, Parent Project Muscular Dystrophy, and Cincinnati's Children Hospital. A cross-sectional design with quantitative methodology was used to examine the relationships among maternal uncertainty, coping efficacy, hope, and other mother and child characteristics. Several open-ended questions were included to assess how mothers appraise uncertainty.

Results: The predominant focus of mothers' uncertainty was medical management and social support. Multivariate analysis revealed that older mothers' age, higher hope scores, and having less ambulatory children were significantly associated with less uncertainty. Mothers with lower hope scores, higher perceptions of uncertainty, and those reporting being less spiritual were less confident in their ability to cope with their child's DMBD.

Conclusions: Because younger mothers and those with ambulatory children with DBMD

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perceive more uncertainty, especially uncertainty related to medical management and social support, efforts to help mothers manage uncertainty may be more effective if tailored towards mothers of children with new diagnosis and specific domains of uncertainties most salient to them. Additionally, hope seems to be a factor in shaping uncertainty appraisals and facilitating coping efficacy. Although future studies are needed, interventions aimed at bolstering maternal hope or guiding mothers with low hope to other uncertainty management and reappraisal strategies may be helpful.

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BACKGROUND

There has been a considerable amount of research focused on the psychological outcomes of family members, especially mothers, who care for a child with a chronic physical condition. These significant and constant caregiving demands can have a negative effect on the caregiver's physical and emotional health (Raina et al., 2004). Adaptation is a multidimensional temporal process of coming to terms with the implications of a health threat and the outcomes of that process (Biesecker and Erby, 2008). Some parents adapt to caregiving demands and the chronic nature of the child's condition better than others, and previous research suggests that psychological adaptation is a significant predictor of well-being among caregivers of children with chronic illness (Samson and Siam, 2008). Therefore, elucidating what factors influence caregivers' ability to adapt could inform clinicians how best to facilitate the adaptation process in these families (Raina et al., 2004).

Across disorders, an important factor that has emerged as an important construct in understanding the impact of a condition on parental adaptation is uncertainty (Stewart and Mishel, 2000). Uncertainty pervades many aspects of the illness experience and can arise from numerous factors, including prognostic and disease-course ambiguity (Han et al., 2011). Research investigating sources of distress in the chronic illness context have described uncertainty as "probably the greatest single psychological stressor" faced by the patient and their families (Koocher, 1984).

One challenging chronic physical condition faced by families includes Duchenne and Becker Muscular Dystrophy (DBMD). Duchenne muscular dystrophy (DMD) is an X-linked recessive neuromuscular disorder characterized by progressive loss of muscle function beginning in childhood and leading to an early death. DMD affects about 1 in

every 3,500 newborn males and is caused by a mutation in the gene that encodes the muscle protein dystrophin (Roland, 2000). Affected boys typically manifest symptoms of muscle weakness around ages 2-3 and require a wheelchair by ages 10-12 (Flanigan, 2014). Current medical management aims at slowing the progression of muscle weakness and improving patients' quality of life, but there is currently no cure for DMD and death often occurs by the time patients reach their early 20s (Eagle et al., 2002). Becker muscular dystrophy (BMD), affecting 1 in every 30,000 males, is generally milder than DMD, with a later onset and less severe muscle weakness (Flanigan, 2014). These conditions together are referred to as Duchenne/Becker muscular dystrophy (DBMD).

Uncertainty and caring for a child with DBMD

DBMD is notable among chronic childhood conditions in that it has been described as a "complex chronic condition," causing challenges exemplified by both chronic and terminal illnesses (Nereo et al., 2003). Psychological adaptation for these families is multifaceted because of the evolution of the disease, changing nature of caregiving, and the continual losses that these children and parents face (Samson et al. 2009).

Contributors to uncertainty have not been studied specifically among parents of children with DBMD, but mothers have reported uncertainty related to what the future will hold for their child (Samson et al., 2009). Although the clinical course of most boys with DBMD follows a pattern of progressive deterioration and ultimately death, prognostic variability in these conditions still exists, especially in relation to the timing of loss of skills and lifespan. Cardiomyopathy is a pervasive issue for these individuals, and when significant cardiac impairment will begin is also difficult to predict (Roland, 2000).

Additionally, respiratory failure, the most common cause of death for individuals with DMD, often occurs unexpectedly due to infection (Wong, 2005). Therefore, it is difficult for healthcare providers to predict with specificity when children with DBMD may die (Parker et al., 2005). Not knowing whether or when a child will die is a hallmark of parental uncertainty in life-threatening conditions (Stewart and Mishel, 2000).

Impact of parental uncertainty

The persistent uncertainty described by parents of children with chronic medical conditions has been linked to several significant outcomes for parents and families, both negative and positive. Research have investigated parents' experience and found that uncertainty negatively affects parental coping (Rosenthal, 2001; Lipinski et al., 2006). Psychological distress, anxiety, depression, and hopelessness have also been associated with parental uncertainty (Stewart and Mishel, 2000). Alternatively, parents have reported benefits to uncertainty, including recognizing that uncertainty allows for the possibility for positive outcomes for their child (Stewart and Mishel, 2000; Rosenthal et al., 2001). This dichotomization of uncertainty appraisals demonstrates that uncertainty is not always a state to be avoided or eliminated and some components of uncertainty can be viewed as a positive. It has been suggested that how an individual appraises the uncertainty they perceive, in part, determines his or her ability to adapt to both the uncertainty and the stressor (Mishel, 1990).

A better understanding of factors that impact uncertainty may lead to targeted interventions that enhance the adaptation process. Hope has been presented as having therapeutic value affecting coping and adaptation in both caregivers and patients (Miller, 2007; Herth, 1989; Bruhn, 1984). Parental hope is a promising factor in assessing how

parents appraise DBMD-related uncertainties as they adapt, but this relationship has yet

to be studied. Since mothers of children with DBMD face a plethora of uncertainties

related to their sons' condition, it is important to examine the relationships between

uncertainty, hope, and coping efficacy.

Conceptual framework

This study aims to explore relationships between perceived parental uncertainty,

hope, and coping efficacy within the context of the DBMD. Relationships between these

concepts are understood in light of an integrated model of uncertainty and hope in

adaptation (Lazarus and Folkman's Transactional Model of Stress and Coping (1984),

Mishel's Uncertainty in Illness Theory (1988), and Dufault and Martocchio's Model of

Hope (1985). Figure 1 depicts the conceptual framework that frames this study.
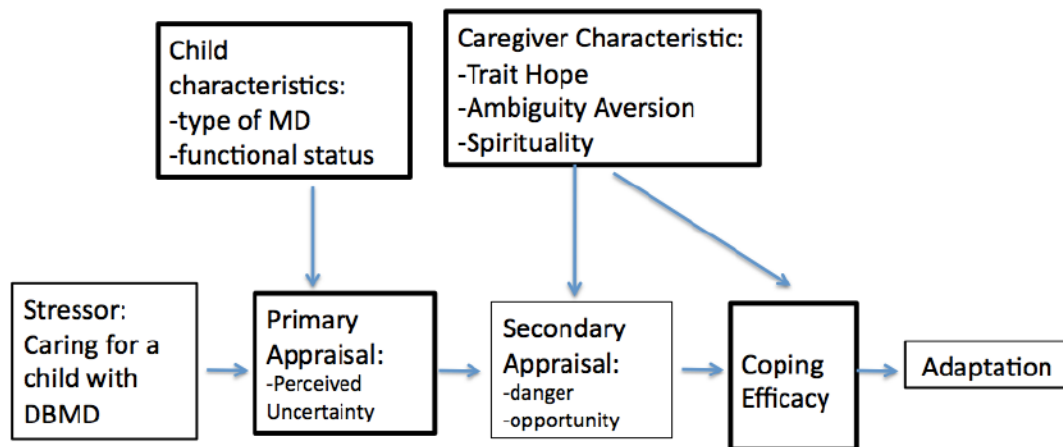
Figure 1. Conceptual Framework: An integrated model of uncertainty and hope in
adaptation

Our integrated model includes key constructs from the Transactional Model of

Stress and Coping (TMSC). The TMSC frames the process of psychological adaptation to

a health stressor, such as a diagnosis of a child with DBMD (1984). Two processes

dictate an individual's response and adaptation to this stress: appraisal and coping

(Folkman and Greer, 2000). The appraisal of the threat is influenced by both an evaluation of its personal significance and an evaluation of what can be done about the stressor. These appraisals determine whether the situation is appraised as a threat or a challenge, directly influence subsequent coping strategies, and thus are important antecedents in adaptation (Folkman and Greer, 2000).

Mishel's Uncertainty in Illness Theory (UIT) further elucidates how parents cognitively process illness related stimuli and how they structure meaning from these events in the context of uncertainty. Mishel defines uncertainty as an "inability to determine the meaning of illness related events or predict outcomes due to a lack of sufficient cues" and includes four domains: ambiguity surrounding the state of the illness, complexity regarding treatment, lack of information about the diagnosis, and unpredictability of the prognosis (Mishel, 1981). This model presents uncertainty as a central feature of the illness experience. Managing uncertainty is an essential task in adaptation (Mishel, 1988). Mishel argued that uncertainty is classified as neutral until it has been appraised as either a danger or opportunity (similar to the TMSC's threat or challenge appraisals). The theory states that when uncertainty is viewed as a danger, an individual will work to decrease the uncertainty. On the other hand, when uncertainty is appraised as an opportunity, the individual will employ coping strategies to foster the uncertainty. Therefore, the ability of an individual to employ effective methods of coping that reflect their appraisal of the uncertainty will impact their adaptation to the stressor (Mishel 1988).

There is diversity in the operationalization of the concept of hope, and no universal definition exists. In health psychology, hope has been found to play a

significant role in the well-being of ill individuals and their caregivers (Miller, 2007). Recognizing the nuances of hope, researchers Dufault and Martocchio defined hope as "a dynamic life force characterized by a confident yet uncertain expectation of achieving a future good which is realistically possible and personally significant" (Dufault and Martocchio, 1985). This definition suggests that hope is a variable that is situation and state-dependent and understood within a context of uncertainty. However, the current available hope scales conceptualize hope as a trait variable (Herth, 1989). We were interested in a conceptualization of situational hope related to DMD to explore the relationships with uncertainty and wellbeing. Therefore, using existing literature on hope and pervious experience through a series of interview studies of parents of children with DBMD, a novel state hope measure was developed with several elements of dynamic situational hope. These include the concepts that hope is grounded in routine experience, exists within uncertainty, and is influenced by relational and spiritual resources (Samson et al., 2009; Duggleby et al., 2010).

Understanding the Relationship between Hope, Uncertainty, and Coping Efficacy

Coping Efficacy

Coping efficacy, also known as coping self-efficacy, is an individual's perceptions of his/her ability to successfully or effectively cope with a given situation. Higher coping efficacy has been identified as a dimension of and antecedent of adaptation (Madeo et al., 2012). Data from the baseline survey of this cohort of mothers with children with DBMD found that mothers with higher coping self-efficacy reported being more highly adapted to their child's condition (Peay, Personal Communication, November 2013).

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Hope and Uncertainty

The concepts of hope and uncertainty are intrinsically linked. A meta-analysis of the hope experience in caregivers of persons with chronic conditions identified that one theme of hope is "dynamic possibilities within uncertainty" (Duggleby et al., 2010). If the future were certain, hope would have no role in the process of expecting a positive future. Therefore, uncertainty is a precondition for hope, as hope is about possibilities. Although there is abundant literature supporting the positive association between hope and well-being, there is little documentation of the role of hope in influencing perceived uncertainty and the uncertainty appraisal process. The only study identified by the researchers found that in a population of cancer survivors, hope was significantly correlated with lower levels of perceived uncertainty (Wonghongkul et al., 2000).

Hope and Positive Psychological Outcomes

Studies examining the relationship between hope and adaptive outcomes have found that hope is negatively correlated with anxiety, dysphoria, and depression (Snyder et al., 2003; Chang and DeSimone, 2001) and positively correlated with quality of life, life satisfaction, and well-being (Bailey et al., 2007; Davis, 2005). Hope is a moderator between disability-related stress and maladjustment in mothers of children with chronic physical conditions, and mothers with high hope are more likely to be better adjusted in the face of disability-related stress than mothers with low hope (Horton and Wallander, 2001). Hope has also frequently been studied in the cancer population and has been found to be positively related to coping style, coping level, and coping effectiveness among adults with cancer (Herth, 1989; Felder, 2004; Zhang et al., 2010). These findings suggest that hope is a significant contributor to the adaptation process.

This study aims to assess how hope influences coping efficacy. It is hypothesized that hope moderates perceptions of uncertainty and directs coping efficacy. Although there are no systematic studies that examine the direct relationship between hope and the uncertainty appraisal process in the DBMD population, Wonghongkul and colleagues have studied the relationship between hope, uncertainty, and stress appraisals in women who have survived breast cancer. They conceptualized hope as a factor that affected stress appraisals and coping and found that hope influenced a positive reappraisal coping strategy (2000). Additionally, Truitt et al. hypothesized that hope is a moderator between parental perceived uncertainty and adaptation in caregivers of children with Down syndrome. The study did not find hope to be a statistically significant moderating variable. However, this cohort perceived relatively low levels of uncertainty, which could have affected this moderation analysis (Truit et al., 2012).

Despite qualitative studies and theoretical considerations suggesting that hope is an important part of the process of appraising and adapting to uncertainty, quantitative data do not yet exist to support this suggestion. The current study will examine the relationship between uncertainty, hope, and coping efficacy among mothers of children with DBMD.

SPECIFIC AIMS AND HYPOTHESIS

Aim 1: To determine if muscular dystrophy type, child's functional status, and maternal hope, spirituality, and ambiguity aversion are associated with maternal perceptions of uncertainty.

> *Hypothesis 2.1: Having children with a Becker muscular dystrophy diagnosis and less mobility will be associated with increased uncertainty.*
>
> *Hypothesis 2.2: Mothers who have lower levels of spirituality, lower levels of hope, and higher levels of ambiguity aversion will be associated with increased uncertainty.*

Aim 2: To assess the relationships among maternal uncertainty, hope, spirituality, ambiguity aversion, and coping efficacy.

> Subaim 1: To determine if hope, spirituality, and ambiguity aversion are independent predictors of coping efficacy.
>
> > *Hypothesis 3.1: Higher maternal hope and spirituality, and lower ambiguity aversion will be positively related to coping efficacy.*
>
> Subaim 2: To determine if hope moderates the relationship between maternal uncertainty and coping efficacy.
>
> > *Hypothesis 3.2: The relationship between maternal weighted uncertainty and coping efficacy will vary by degree of hope.*

Aim 3: To describe what mothers report as being most uncertain about having a child with DBMD and how they appraise this uncertainty.

METHODS

Study Population and Recruitment Strategies

This study was nested in a longitudinal 5-year study sending yearly surveys to mothers of children with DBMD. Recruitment was originally conducted through the Duchenne Connect registry, Parent Project Muscular Dystrophy (PPMD), and Cincinnati Children's Hospital Medical Center Neuromuscular Clinic. Individuals were eligible to participate if they were the biological mothers of a living child with Duchenne or Becker muscular dystrophy living in the United States, are 18 years of age or older, and able to answer a survey in English. There was no age requirement for the child with DBMD. Participants were asked to disclose their child's diagnosis but no screening evaluation was required. Additional participants were recruited through the Duchenne Connect registry, and clinicaltrials.gov website. The sample size calculation indicated that 200 participants were needed to have 80% power to detect the effect of a key independent variable explaining at least 5% of the total variance in coping efficacy.

Procedures

This study involved a one-time self-administered survey. Previously recruited participants were sent a pre-survey announcement email about 4 weeks before the survey was sent out (Appendix A). When the survey was opened they were emailed a personalized link to the survey and instructed to either access the electronic version of the survey through SurveyMonkey, a secured Internet site, or to contact the researcher for a paper copy of the survey.

To recruit new participants, a study description (Appendix B) was sent out to Duchenne Connect registry participants and posted on the clinicaltrials.gov website.

Interested individuals were instructed to contact the study investigators (MB or HP) to receive a personalized link to the electronic version of the survey or be sent a paper copy, which was done once the survey was opened. At the beginning the survey a study description provided an overview of the study to ensure that participants were eligible and able to provide consent. Participants were informed that they could withdraw from the study at any time (up until their submission of the survey), that they could skip any question(s), and that they could discontinue taking the survey at any time. Participants provided informed consent by checking a box on the first page of the survey.

The survey was open from September 6, 2014 to November 4, 2014. Individuals who completed the survey received a $20 Amazon.com gift card emailed to them.

This study was approved by the National Human Genome Research Institute's Institutional Review Board (Protocol # T-HG-0108) and the Cincinnati Children's Hospital Institutional Review Board.

Study Design and Instrument

This study used a cross-sectional research design with a quantitative survey that included open-ended questions. Validated instruments were used to assess coping efficacy, maternal uncertainty, spirituality, and ambiguity aversion. The hope scale used was purposely developed for this study. The parental uncertainty of children's health scale, hope scale, and open-ended questions were piloted among ten mothers of children with DBMD known to the co-investigator (HP) and changes were made as needed to clarify the meaning of several items and to better define the prompts.

Demographic information: Mother and Child with DMBD

Demographic information on returning participants was compiled from the

baseline survey. Mothers were asked to update demographic information about themselves including: their employment status, marital status, annual income, whether they have had any children in the last 12 months (and whether that child has DBMD), and whether any of the children have died in the last 12 months (and whether that child had DBMD). Additionally, they were asked to update demographic information about their child with DBMD including: the functional status of their child with DMBD (a 7-item categorization focused on ability to ambulate, which includes arm functionality for non-ambulatory individuals, as used in the Duchenne Connect Registry, www.duchenneconnect.org), and the child's previous participation in a clinical trial.

New participants were asked to provide information about their income, age, gender, race/ethnicity, marital status, level of education, employment, number of children with DBMD, number of children, and maternal carrier status. Information was also collected about their child with DMBD, including the child's diagnosis (Duchenne, Becker, or intermediate phenotype), current age, age at diagnosis, gender, functional status (as described above), and the child's previous participation in a clinical trial.

Maternal Uncertainty

Maternal uncertainty was measured using a revised version of the Parental Uncertainty of Children's Health Scale (PUCHS), a scale developed by Biesecker and colleagues (Macnamera et al., 2014). The PUCHS has been used previously in one study of caregivers of children with undiagnosed conditions and includes items targeting four dimensions of maternal uncertainty: medical management (items 1, 2, 3, 4, 5, 8, 9), reproductive (items 6, 7), social (items 10, 11), and existential (items 12, 13). This 26-question scale is composed of two sections, each with 13 questions. In the first section,

participants were asked to rank, on a 5-point scale, the level of uncertainty they perceive about two statements representing each dimension of uncertainty. In the second section, participants were asked to rank the level of importance of resolving uncertainty for each of the statements. Each uncertainty item on the PUCHS represents uncertainty by a 5-point Likert-format scale ranging from -2 (strongly disagree) to 2 (strongly agree). Each importance item on the PUCHS represents importance by a 5-point Likert-format scale ranging from 1 (unimportant) to 5 (most important).

A weighted uncertainty score was calculated for each of the dimensions using the following formula, with U=uncertainty and I=importance:

Reproductive Weighted Uncertainty $= ((U_6 \times I_6) + (U_7 \times I_7)) / (I_6 + I_7)$

To determine the total weighted uncertainty score, each question's level of uncertainty was weighed by the importance ranked by the participant, as calculated by the following formula:

Weighted Uncertainty $= [(U_1 \times I_1) + (U_2 \times I_2) + ... + (U_{13} \times I_{13})] / [(I_1 + I_2 + ... + I_{13})]$

The alpha internal consistency coefficients for the medical management, reproductive, social, and existential dimensions were 0.84, 0.62, 0.81 and 0.94, respectively. The reliability coefficient of the PUCHS has been previously calculated as 0.79 (Macnamara et al., 2014) and the reliability coefficient of the scale in this study was calculated as 0.76.

Hope

A novel hope scale was created by co-investigator Holly Peay to measure situational disorder-specific hope. It is a 12-item measure asking mothers to respond to how often they have found hope for the future in the following domains: hope grounded

in routine experience, hope grounded in a sense of personal expertise, hope grounded in the present, hope grounded in resources, optimistic orientation, the promise of uncertainty, and acceptance with hope. This scale uses a 1-5 Likert scale anchored by "Never" to "Always." This scale has not previously been used and a principle components analysis was conducted to examine the number of domains, followed by reliability analysis.

Coping Efficacy

The Coping Self-Efficacy Scale (CSES) was used to measure a person's perceived ability to cope effectively with a stressor (Chesney, 2006). It is a 26-item measure that uses a 0-10 scale anchored by "cannot do at all" and "certain can do." An overall score is created by summing the item ratings and higher scores indicate greater coping self-efficacy. This scale has been shown to be valid and reliable (Cronbach's $\alpha = 0.80$-0.91) and has previously been used in a study of caregivers of patients with cancer (Mosher, 2013). The reliability coefficient of the scale in this study was 0.96.

Spirituality

The Daily Spiritual Experience Scale (DSES) is a 16-item self-report measure of spiritual experience. The first 15 items of the questionnaire are measured on a 6-point Likert-type scale: many times a day, every day, most days, some days, once in a while, and never or almost never. Item 16 is measured on a 4-point scale: Not Close at All, Somewhat Close, Very Close, As Close as Possible. Scores are then summed for a total composite DSES score for each individual and ranged from 16 to 94, with higher score indicating less spiritual experience (Underwood and Teresi, 2002). This scale has been used in many studies in the medical field, including one analyzing the relationship between chronic illness and psychological well-being (Ballew et al., 2001). The

reliability coefficient of the scale in this study was calculated as 0.97.

Ambiguity Aversion

The Ambiguity Aversion Scale was used to assess the content validity of the maternal uncertainty PUCHS scale. Ambiguity is a specific type of uncertainty that results from lack of reliability, credibility, or adequacy in information (Han et al., 2011). Ambiguity aversion, or intolerance of ambiguity, is a response to this ambiguity. This 6-question scale is measured on a 5-point Likert scale (anchored by "strongly disagree" to "strongly agree") and has been used in populations including physicians and medical students but not in parents or caregivers (Politi et al., 2011; Geller, 2013). The reliability coefficient of the scale in this study was 0.72.

Open-Ended Questions: A total of five open-ended questions were included as part of this study Because the uncertainty measure has not yet been used in this population, and there is currently no method for measuring how the participants are appraising uncertainty. Therefore, four free-response questions were included to better assess what mothers' perceive as most uncertain, how this uncertainty has affected them, and how they appraise the uncertainty. Responses to two of these questions were qualitatively and quantitatively explored. Additionally, one open-ended question asked mothers about how their hope has changed over time, which also helped us evaluate the new hope measure.

Data Analysis

Data were analyzed using STATA 13.1. Most of the demographic variables with multiple response categories or continuous responses were dichotomized after performing preliminary descriptive analyses. These included: annual income (<$99,999 vs. >

$100,000), marital status (not married/long-term relationship vs. married/long-term relationship), level of education (completed college or more vs. some college or less), race (Caucasian vs. not), employment (not employed vs. full time/part time employed), number of children with DBMD (1 vs. >1), and maternal carrier status (carrier vs. not carrier/do not know carrier status), and the child's DBMD diagnosis (Duchenne vs. Becker/intermediate phenotype). The following variables were categorical: number of children (1 vs. 2 vs. 3 vs. >3), oldest affected child's age at diagnosis (prenatal vs. 0-3 years vs. 4-7 years vs. 8-11 years vs. 12 years or older), and oldest affected child's functional status (ambulatory children (presymptomatic/early ambulatory) vs. children in transition to the use of a power wheelchair full-time (late-ambulatory/early non-ambulatory) vs. full-time users of power wheelchairs (non-ambulatory 1-III)). Key predictor variables (uncertainty, hope, spiritualty, ambiguity aversion, and coping efficacy) remained continuous variables in the analysis. Participants were allowed to skip questions, and if greater than 80% of the scale was completed, missing values were filled by replacing the missing value with an average of all the other items. If less than 80% of a scale was complete, the variable was dropped. To keep the sample size consistent, any participant with a dropped key or demographic variable was eliminated from the data set.

Descriptive analyses were performed on all numerical variables. T-tests and chi-square tests were used to compare key variables and other covariates between the returning participants and newly recruited participants. Bivariate exploratory analysis was conducted among the key predictor variables (hope, uncertainty, coping efficacy, ambiguity aversion, spirituality, child's functional status, and MD type), as well as

between uncertainty and coping efficacy and potential confounding variables (other child and mother characteristics and demographic variables) using Pearson's and Spearman's correlations. All independent variables that had a significant bivariate relationship with uncertainty at the $p<0.05$ level were regressed on to uncertainty. A backwards elimination strategy was used to remove one variable at a time, starting with the highest p-value, until only those with a p-value of <0.05 remain. A change in estimate strategy was used to assess the impact of confounding variables. Each potential confounder was added to the model individually and if the beta for any key predictor variables in the model changed by more than 10%, the confounder was included in the final model.

For analysis of the open-ended questions, 3 of the 5 free questions were coded (questions 1, 2 and 5). A codebook was developed for each question through thematic analysis. Themes were informed by common maternal responses as well as the Transactional Model of Stress and Coping, as uncertainty appraisals are often described as a threat or opportunity. For question 1, each response was labeled with up to 3 categories from the codebook. For questions 2 and 5, each response was label with up to 2 categories from the codebook. The number of responses within each category were quantified and presented descriptively.

RESULTS

Response

Returning participants

During the recruitment period from September 6, 2014 to November 4, 2014, emails were sent to 200 mothers who had previously consented to be part of the longitudinal study and completed the baseline survey. Of these, 8 of the emails "bounced back," 147 participants started the survey, and 144 finished the survey. In the 2.0% (n=3) of incomplete surveys the participant did not answer the survey beyond the open-ended questions and, because more than half of the survey was not answered, these responses were not included in the analysis. There was a 76.5% (147/192) response rate, similar to the first year follow-up survey that had a response rate of 78%.

New participants

One hundred and twelve individuals contacted the study investigators with interest in joining the study. Ninety-two of these individuals started the survey, but 3 (3.2%) were incomplete and 5 (5.4%) did not meet the eligibility criteria.

Overall there were 228 completed surveys by eligible mothers. If a scale was less than 80% completed or a key variable or potential confounding variable was missing, the entire entry was eliminated from the data set to keep the sample size consistent throughout analyses. One response was eliminated due to an incomplete uncertainty scale, 1 due to an incomplete hope scale, one due to an incomplete spirituality scale, one due to unanswered clinical trial participation, and three due to an unanswered maternal age question. Therefore, the final sample size for data analysis was 221.

Demographics of Participants and Participants' Children

The mean age of the mothers was 44.9 (± 8.9), with a range of 26 to 68. One hundred and eighty-seven (84.6%) mothers were married or in a long-term committed relationship, 27 (12.2%) were divorced or separated, 4 (1.8%) had never married, and 3 (1.4%) were widowed. Two hundred and three mothers identified as being Caucasian (91.9%), 6 (2.7%) as Asian, 14 (6.4%) as Hispanic, 2 (0.9%) as African American, and 14 (6.3%) as Other; respondents had the option of endorsing more than one category. The majority of participants had completed college or post-graduate studies (72.4%) and was employed or attended school part- or full-time (69.2%). The median household income was $50,000-$99,999. One hundred participants (45.3%) were carriers of mutations related to DBMD, 78 (35.3%) were non- carriers, and 43 (19.5%) did not know their carrier status.

Two hundred and five mothers had one affected child (92.8%) and 16 (7.3%) had two or more affected children. The majority of the oldest affected children had DMD (187, 84.6%), 24 (10.9%) had BMD, and 10 (4.5%) had an intermediate phenotype. The mean age of the participants' oldest child with DBMD was 14.1 ± 7.3 years with a range of 2 years to 37 years old. Two children were diagnosed prenatally, 86 (39.8%) between 0-3 years, 109 (49.3%) between 4-7 years, 21 (9.5%) between 8-12 years, and 3 (1.4%) after 12 years of age.

The mean child functional status was 3.7 (±1.8) with higher numbers indicating less ambulation. The 7-item child functional categorization was re-coded into three ambulation categories: ambulatory children (85, 38.5%), children in transition to the use of a power wheelchair full-time (43, 19.4%), and full-time users of power wheelchairs

(93, 42.1%). If the participant had more than one affected child, the functional status, age and age of diagnosis of the oldest living child were reported. Table 1 summarizes the characteristics of the sample.

Table 1: Demographic Characteristics of Study Population

Demographic Characteristic		N	%
Race*	Caucasian	203	91.86
	Asian American/Pacific Islander	6	2.71
	African American	2	0.90
	Native American	0	0
	Other	14	6.33
Ethnicity	Not Hispanic or Latino	206	93.64
	Hispanic or Latino	14	6.36
Current Marital Status	Single/Never Married	4	1.81
	Married or long-term committed relationship	187	84.62
	Divorced	19	8.60
	Separated	8	3.62
	Widowed	3	1.36
Highest Level of Education	Grade school or middle school	0	0
	High School/GED	11	4.98
	Some College/Technical School	50	22.62
	Completed College	100	45.25
	Post-Baccalaureate Degree	60	27.15
Annual Income	Under $50,000	35	16.13
	$50,000 - $99,999	88	40.55
	$100,000 - $149,999	43	19.82
	$150,000 - $199,999	30	13.82
	$200,000 - $249,999	5	2.30
	Above $250,000	16	7.37
Employment Level of Mother	All of my time is spent caring for my family and my house	68	30.77
	Part-time job or school	108	48.87
	Full-time job or school	45	20.36
Carrier Status of Mother	Carrier	100	45.25
	Not a Carrier	78	35.29
	Don't know	43	19.46
Gender of Child with DBMD	Male	220	99.55
	Female	1	0.45
Diagnosis of Child with DBMD	Duchenne	187	84.62
	Becker	24	10.86
	Intermediate Phenotype	10	4.52
Age at Diagnosis	Prenatally	2	0.90

of Oldest child with DBMD	0-3 years		86	38.91
	4-7 years		109	49.32
	8-12 years		21	9.50
	>12 years		3	1.36
Functional Status of Oldest Child with DBMD	Presymptomatic		17	7.69
	Early-ambulatory		68	30.77
	Late-ambulatory		34	15.38
	Early non-ambulatory		9	4.07
	Non-ambulatory I		50	22.62
	Non-ambulatory II		32	14.48
	Non-ambulatory III		11	4.98
Current Age of Oldest Child with DMBD	≤4		8	3.62
	5-9		59	26.70
	10-14		62	28.05
	15-18		35	15.84
	19-22		26	11.76
	≥23		31	14.03

Descriptive Analysis

Maternal Uncertainty

The PUCHS was used to assess parents' perceptions of uncertainty and the related importance of resolving the uncertainty. Total uncertainty scores could range from -2 to 2, with higher scores indicating that an individual perceived more uncertainty about areas of importance to them related to their child's DBMD. Scores for total uncertainty ranged from -2 to 1.48, and the mean was -0.71 ± 0.78.

Dimensions of Perceived Uncertainty

There are four distinct dimensions of uncertainty identified in the PUCHS: medical management, reproductive, social, and existential. Participant's weighted medical management uncertainty scores ranged from -2 to 2, and the mean was -0.56± 0.87. The weighted reproductive uncertainty scores ranged from -2.00 to 2.00, and the mean was -1.03 ± 1.05. The weighted social uncertainty scores ranged from -2.00 to

2.00, and the mean was -0.77 ± 1.11. The weighted existential uncertainty scores ranged

from -2.00 to 2.00, and the mean was -1.04 ± 1.13.

Coping Self-Efficacy

Table 3 presents the mean scores and standard deviations for the participants'

responses to the Coping Self-Efficacy Scale, with higher scores indicate greater

confidence in one's ability to cope. The coping self-efficacy scores ranged from 14-260,

with the mean being 166.97 ± 47.35.

Hope

A Principle Components Analysis (PCA) indicated that there was one domain

across items 1-10 and 13-16. Due to differing stems, questions 11 and 12 were left out of

the components analysis to be analyzed separately (data not shown).

Table 2: Factor Analysis of Hope Scale

In the past 4 weeks, how often have you found hope in...	Components
	1
1) ... The day-to-day life with your child	.688
2) ... Aspects of your child's medical care	.775
3) ... Your confidence about your child's future	.733
4) ... Your areas of expertise and know-how in caring for your child	.517
5)... Your child's long-term health could be better than expected	.746
6)... Research being done today	.648
7)... The uncertain potential of your child	.697
8)... Understanding your child	.639
9)... Imagining your child's bright future	.790
10)... Parenting your child	.646
In the past 4 weeks, how often have you found hope through...	
13)... Your use of the resources available to you	.654
14)... Your determination	.688
15)... Your faith or spirituality	.602
16)... Your use of the resources available to you	.594

The hope score was calculated as an average of the 14 questions, with a higher score indicating higher hope. The range of average hope scores was 1.8 – 5 and the mean hope score was 3.70 ± 0.62.

Spirituality

The spirituality score was calculated by summing the scores of the scale, with a higher score indicating lower reported spirituality. The range of spiritualty scores was 16-93 with a mean score of 49.25 ± 19.88.

Ambiguity Aversion

The ambiguity aversion score was calculated by averaging the 6 scores, with a higher score indicating more aversion to ambiguity. The range of ambiguity aversion scores were 1.33 – 4.50 with a mean score of 3.09 ± 0.51

Table 3. Descriptive Data of Key Variables

N=221	Mean ± SD	Range of Study Sample	Range of Possible Scores
Total Weighted Uncertainty	-0.71 ± 0.78	(-2 – 1.48)	(-2 – 2)
Medical Management Uncertainty	-0.56 ±0.87	(-2 – 2)	(-2 – 2)
Reproductive Uncertainty	-1.03 ±1.05	(-2 – 2)	(-2 – 2)
Social Uncertainty	-0.77 ±1.12	(-2 – 2)	(-2 – 2)
Existential Uncertainty	-1.05 ±1.13	(-2 – 2)	(-2 – 2)
Coping Self-Efficacy	166.97 ± 47.35	(14 – 260)	(0 – 260)
Hope	3.70 ± 0.62	(1.8 – 5)	(1-5)
Spirituality	49.25 ± 19.88	(16 – 93)	(16-94)
Ambiguity Aversion	3.09 ± 0.51	(1.33 – 4.5)	(1-6)

Comparison of Returning Participant Sample to New Participant Sample

The participants from this study were recruited at two different time points. One hundred and thirty-nine participants had been previously recruited to be part of a 5-year longitudinal study on predictors of the wellbeing of mothers of children with DBMD. The 5-year study includes yearly surveys of which this survey was part of year 3. Eighty-two participants were newly recruited to this longitudinal study, and this was their first survey. These groups were recruited about 3 years apart. If the recruitment sources had similar age distributions between the two recruitment times, then it was likely that the new participants would have younger, more ambulatory children.

Therefore, t-tests and chi-squared tests were performed to compare the levels of key predictor and demographic variables. Table 4 presents the differences and p-values of the t-tests between the two groups for the continuous key predictor variables. Children of the new participants, as well as the new participants themselves, were significantly younger than the current age of the returning participants and their children. In addition, there was a significant difference in the total weighted uncertainty level between the returning and new participants, with the returning participants having a more negative uncertainty score (less uncertainty).

Table 4: T-Test Comparing Returning and New Participant Groups

| | Group | | |
	Returning Participants (SE) n=139	New Participants (SE) n=82	Difference
Uncertainty	-0.819 (0.062)	-0.533 (0.091)	-0.286**
Hope	3.706 (0.054)	3.694 (0.066)	0.012
Spirituality	48.629(1.690)	50.318 (2.194)	-1.689
Ambiguity Aversion	3.060 (0.046)	3.136 (0.049)	-0.076
Age of child	15.158 (0.621)	12.317 (0.762)	2.841**
Mom's age	46.295(0.721)	42.460 (1.004)	3.835***
Functional Status	3.820 (0.152)	3.402 (0.198)	0.418*

* $p<0.05$ level, **$p<0.01$, ***$p<0.001$

Table 5 presents the chi-square analysis of the differences in dichotomous variables between the 2 groups. The results indicated that there were no statistically significant differences between the two groups for the following variables: marital status, race, ethnicity, education level, annual income, employment status, carrier status, diagnosis, and age at diagnosis.

Table 5: Chi-Square Comparing Returning and New Participant Groups

| | | Group | |
		Returning Participants	New Participants
Marital Status	Married	118	69
	Not Married	21	13
Race	Caucasian	128	75
	Not Caucasian	11	7
Ethnicity	Not Hispanic	130	76
	Hispanic	9	5
Education	Some college or less	37	24
	Completed college or more	102	58
Annual Income	≤ $99,999	74	49
	≥ $100,000	62	32
Employment	Not working	43	25
	Full/Part-time job or school	96	57
Carrier Status	Carrier	65	35
	Not a Carrier/Do not know	74	47
Diagnosis	DMD	114	73
	BMD/intermediate	25	9
Age at Diagnosis	Prenatally	2	0
	0-3 years	58	28
	4-7 years	66	43
	8-12 years	10	11
	>12 years	3	0

*p<0.05 level, **p<0.01, ***p<0.001

Aim 1: To determine whether muscular dystrophy type, child's functional status, spirituality, and ambiguity aversion are associated with maternal perceptions of uncertainty.

Bivariate Analysis

Using the conceptual model (Figure 1) as a framework for understanding relationships among key variables, bivariate analysis was performed to determine the strength and significance of predicted relationships. Bivariate analyses revealed significant (p<0.05) associations between total weighted uncertainty and hope, spirituality, ambiguity aversion, and child's functional status (See Table 6).

Table 6: Correlations Among Key Variables

N=221	Coping Efficacy	Total Uncert.	Hope	Spirit.	Ambig. Aver.	Child's funct. status	DMBD Dx
Coping Efficacy	1.000						
Total Uncertainty	-0.436***	1.000					
Hope	0.567***	-0.325***	1.000				
Spirituality	-0.458***	0.238**	-0.502***	1.000			
Ambiguity Aversion	-0.073	0.140*	-0.099	0.107	1.000		
Child's functional status	0.023	-0.265**	0.015	-0.003	-0.076	1.000	
DBMD Diagnosis	0.009	0.113	0.059	0.000	-0.001	-0.269**	1.000

*p<0.05 level, **p<0.01, ***p<0.001

Additionally, bivariate analyses were conducted between total weighted uncertainty and all demographic variables. Table 7 depicts the correlations between uncertainty and dimensions of uncertainty with child characteristics. Both clinical trial participation and child's age were statistically correlated with less maternal uncertainty. This relationship was similar for medical management uncertainty, as well as social uncertainty. Reproductive uncertainty was negatively correlated with child's age, and existential uncertainty was not significantly correlated to any child characteristic.

Table 7: Correlations Between Uncertainty and Child Characteristics

N=221	Clinical Trial	Age at diagnosis	Child's age
Uncertainty Total	**-0.214****	-0.025	**-0.241*****
Uncertainty -med man.	**-0.197****	-0.030	**-0.250*****
Uncertainty -reproduct.	-0.036	-0.080	**-0.221****
Uncertainty -social	**-0.211****	0.064	**-0.142***
Uncertainty -existential	-0.103	0.015	-0.049

*p<0.05 level, **p<0.01, ***p<0.001

Pearson's and Spearman's correlations between uncertainty and dimensions of uncertainty with mother's demographic variables indicated that mothers who were older, carried the DBMD mutation, or had a greater number of affected children had statistically lower uncertainty levels (Table 8). Carrier status had a significant positive association with medical management uncertainty and reproductive uncertainty. A younger age of mothers was significantly associated with more medical management and reproductive uncertainty. Having a greater number of affected children was significantly associated with only medical management uncertainty and existential uncertainty.

Table 8: Correlations between Uncertainty and Mother Characteristics

N=221	Carrier status	Mom's age	Income	Edu. level	Marriage status	Employ -ment	# kids affected
Uncertainty	**0.167***	**-0.236****	0.035	-0.035	-0.006	-0.045	**-0.160***
Uncertainty -med man.	**0.141***	**-0.249****	0.006	-0.027	0.003	-0.025	**-0.149***
Uncertainty -reproduct.	**0.173****	**-0.252****	-0.071	-0.080	-0.015	-0.026	-0.041
Uncertainty -social	0.077	-0.091	0.056	0.013	-0.051	-0.001	-0.069
Uncertainty -existential	0.229	-0.062	0.063	0.011	-0.072	-0.030	**-0.173***

*p<0.05 level, **p<0.01, ***p<0.001

Multivariate Analyses

Since uncertainty was significantly associated with hope, spirituality, ambiguity aversion, child's functional status, and the child's DBMD diagnosis in bivariate analysis, we examined the strength of these relationships while controlling for the other key variables and potential confounders using a multivariate regression though a backwards elimination strategy. The final uncertainty regression included hope, child's functional status, and mother's age (Table 9). Older mothers' age (p=0.001), higher hope scores (p<0.001), and having less ambulatory children (p=0.018) were associated with less uncertainty.

Table 9: Final Multilinear Regression with Uncertainty as the Dependent Variable

Model (R^2=0.2169) N=221	Unstandardized Coefficients		Standardized Coefficients	t	P-value
	Beta	Std. Error	Beta		
Hope	**-0.459**	**0.077**	**-0.366**	**-5.95**	**<0.001*****
Functional status	**-0.141**	**0.059**	**-0.162**	**-2.39**	**0.018***
Mom's age	**-0.020**	**0.006**	**-0.230**	**-3.31**	**0.001****
Constant	2.170	0.408		5.32	<0.001

*p<0.05 level, **p<0.01, ***p<0.001

The R^2 for the regression model shown in Table 9 was 0.2169, indicating that these 3 variables together accounted for 21.69% of the variance in total weighted uncertainty. The standardized beta coefficients estimate effect size, with hope having the largest effect size, followed by mother's age and child's functional status. Based on the undstandardized Beta Coefficient results, For every 1 point increase in hope, there is a 0.459 decrease in total uncertainty, when controlling for child's functional status and mother's age. For every 1 point increase functional status, mothers have 0.141 less uncertainty when controlling for hope and mother's age. For every year increase in their age, mothers have a 0.020 decrease in uncertainty.

Aim 2: To assess the relationships among maternal weighted uncertainty, hope, spirituality, ambiguity aversion, and coping efficacy.

Bivariate Analysis

Bivariate analyses of the key predictor variables revealed significant (p<0.05) associations between coping efficacy and uncertainty, hope, spirituality (Table 6). Additionally, bivariate analyses were conducted between coping efficacy and all demographic variables. There were no statistically significant correlations between

coping efficacy and child characteristics and mother characteristics, respectively (Table 10 and 11).

Table 10: Correlations Between Coping Efficacy and Child Characteristics

N=221	Clinical Trial	Age at diagnosis	Child's age
Coping Efficacy	0.069	-0.042	0.017

*p<0.05 level, **p<0.01, ***p<0.001

Table 11: Pearson's Correlations Between Coping Efficacy and Mother Characteristics

N=221	Carrier status	Mom age	Annual income	Edu. level	Marriage status	Employ-ment	# kids affected
Coping Efficacy	-0.059	-0.002	-0.040	0.027	0.096	-0.024	0.121

*p<0.05 level, **p<0.01, ***p<0.001

Multivariate Analyses

Since coping efficacy was significantly associated with hope, spirituality, and uncertainty in bivariate analysis, we examined the strength of these relationships while controlling for key variables and potential confounders using multivariate regression through backwards elimination. The final coping efficacy regression included hope, uncertainty, and spirituality (Table 12). Mothers with lower hope scores (p<0.001), higher perceptions of uncertainty (p<0.001), and those reporting being less spiritual (p=0.001) were less confident in their ability to cope with their child's DMBD.

Table 12: Final Multilinear Regression of Coping Efficacy as the Dependent Variable

Model (R²=0.4271) (N=221)	Unstandardized Coefficients		Standardized Coefficients	t	P-value
	B	Std. Error	Beta		
Hope	**28.742**	**4.680**	**0.377**	**6.14**	**<0.001*****
Uncertainty	**-16.085**	**3.324**	**-0.265**	**-4.84**	**<0.001*****
Spirituality	**-0.491**	**0.142**	**-0.206**	**-3.45**	**0.001****
Constant	73.320	21.354		3.43	0.001

*p<0.05 level, **p<0.01, ***p<0.001

The R^2 for the regression model shown in table x was 0.4271, indicating that these 3 variables together accounted for 42.71% of the variance in coping efficacy. Hope had the largest effect size, followed by uncertainty and spirituality. For every 1 point increase in hope, there is a 28.742 increase in coping efficacy when controlling for uncertainty and spirituality. For every 1 point increase in uncertainty, mothers have 16.085 less uncertainty when controlling for hope and spirituality. For each 1 point increase in spiritualty (less spiritual) mothers have a 0.491 decrease in coping efficacy when holding hope and uncertainty constant.

Moderation Analysis

In order to test the hypothesis that the relationship between uncertainty and coping efficacy will vary by degree of hope, an interaction term, hope*uncertainty, was created by multiplying the hope and uncertainty scores. When regressed on coping efficacy along with the main effects of uncertainty, hope, and spirituality, the interaction variable was found to be non-significant (p=0.277), indicating that the relationship between uncertainty and coping efficacy did not significantly vary by degree of hope.

Table 13: Final Moderation Regression of Uncertainty*Hope with Coping Efficacy as the Dependent Variable

Model (R^2=0.4355) (N=221)	Unstandardized Coefficients		Standardized Coefficients	t	P-value
	Beta	Std. Error	Beta		
Hope	**31.227**	**5.262**	**-0.411**	**5.93**	**<0.001***
Spirituality	**-0.495**	**0.143**	**-0.208**	**-3.46**	**0.001****
Uncertainty	**-34.857**	**17.362**	**-0.557**	**-2.01**	**0.046***
Hope*Uncertainty	4.959	4.552	0.326	1.09	0.277
Constant	64.618	22.719		2.84	0.005

*p<0.05 level, **p<0.01, ***p<0.001

Open-Ended Responses about Uncertainty

<u>Aim 3</u>: To describe what mothers report as most uncertain about having a child with DBMD and how they appraise this uncertainty

Question 1: Many parents explain that they feel uncertainty about the future of their child with DBMD. What is one thing about your child's future that feels the most uncertain?

Two-hundred and nineteen mothers provided responses to this free response question. Responses varied in length from a word or a statement to several sentences. Because of the length and depth of some responses, each could be coded with up to three categories. The types of greatest uncertainty that mothers reported tended to fall into 4 major categories: child's health, child life experience, medical care, and caring/mothering. These topics were further broken down into subcategories as shown in Table 13.

Table 14. Quantitative Analysis of Open-Ended Question 1

What feels the most uncertain?	n
Child's Health	**131**
Prognosis	47
Lifespan	63
General health	13
Emotional health	8
Child's Life Experience	**85**
Quality of life	27
Education decisions	16
Independence	14
Employment	12
Marriage/partner	9
Social	7
Caring/Mothering	**23**
Long-term care/after parents pass away	10
Meeting child's needs medically	6
Logistics (home-set up, financial)	5
Explaining DMD to child	2
Medical Care	**19**
Future drugs/treatment	16
Best treatment now	2
Medical team	1

Child's Health

The most common uncertainty response among mothers pertained to their child's

health. Sixty-three mothers indicated that their child's lifespan was the most uncertain

for them and 47 said that their child's functional status prognosis was the most uncertain.

One mother explained uncertainty concerning the circumstances of her son's passing.

121: "Life expectancy as well as when and how to know when the end is near.

Not knowing what that looks like is terrifying. I have not been able to even

visualize this."

Another mother of with a child with BMD explained how the variability of this

form of MD impacted the uncertainty she experiences.

268: "The progression of his symptoms and how long he will be ambulatory, etc. He has BMD and the symptom progression can progress at widely varying rates"

Other uncertainties within the child's health theme included uncertainty about the child's heart health, future medical complications, and emotional health. One mother explained her uncertainty about her son's emotional experience.

371: "Will he suffer or be scared."

Child's Life Experience

Eighty-five responses fell within the theme of uncertainty about the child's life experience, with the most prevalent response indicating uncertainty about the child's quality of life. In their responses, mothers articulated the challenge of how to best use the time that their child has, and how to help their child live a fulfilling, meaningful life.

126: "Not knowing how to guide him to live a full life with a career and family of his own considering his life expectancy and the physical limitations he will endure."

218: "[My child] living long yet having no purpose."

Mothers also had uncertainty relating to if and how their child would live a normal childhood and young adult life including having friends, going to school, attending college, being employed, and getting married.

305: "His relationship with others. He has very few true friends."

118: "Setting expectations for his future; he wants to know what he will be able to do & how he can be successful in work, travel, family"

196: "His ability to find a job after high school."

Caring/Mothering

Twenty-three mothers cited the caring and mothering of their child as uncertainty provoking. Several mothers mentioned more logistical uncertainties including when and how to set up a handicap accessible home and how to financially providing for medical expenses.

111: "When we are going to need to do home modifications or move to a ranch house"

Ten mothers cited uncertainty and worry about how her child would be cared and provided for if she passes away before her child. Additionally, two mothers explained that they are uncertain of the best way to explain and communicate the DBMD diagnosis to her child. Six mothers explained uncertainty stemming from her ability to meet her child's needs.

191: "Could I be doing a better job?"

Medical Care

Nineteen mothers cited their child's medical care as their greatest point of uncertainty. A majority cite the uncertainty of potential new treatments in the future as most uncertain whereas only three mothers cite choosing the best current treatment for their child as most uncertain.

426: "When the drug he needs will be available to him - we need ataluren now and it is available in Europe and every day since he turned 5 is a day lost."

Only one mother citied the medical care team and where to find appropriate care for her son as most uncertain.

223: "Where to obtain good care as he ages. The doctors do not seem to care about his future because he is so much older than everyone else and they think he should already be dead."

Question 2: How has this uncertainty affected your life?

Two-hundred and eighteen mothers provided responses to this free response question. Responses varied in length from a word or a statement to several sentences. Because of the length, depth, and common themes of the responses, each could be coded with up to two categories. Several of the responses did not answer the prompt and were not coded. The common themes for the effects of uncertainty on mothers' lives fell into 7 major categories: emotional, personal, planning, mothering, family-related, practical and financial, and intrusive thoughts. These topics were further broken down into subcategories as shown on Table 15.

Table 15. Quantitative Analysis of Open-Ended Question 2

How has this uncertainty affected your life?	n
Emotional	**83**
Negative impact	76
Positive impact	7
Planning	**50**
Make a plan for the future	9
Take it day to day/do things now instead of waiting	24
Hard to make plans for the future	17
Intrusive Thoughts	
Think about if often	18
Effects every aspect of life	7
Doesn't affect me	21
Practical and Financial	**16**
Home	8
Finances	8
Mothering	**17**
Personal	**14**
Faith	6
Health	3
Career	5
Family	**9**
Marriage	4
Family functioning	5

Emotional

The most frequent response to the effect of uncertainty pertained to the negative emotional burden of the uncertainty. Mothers explained that this uncertainty has caused depression, anxiety, fear, hopelessness, worry, sadness, anger and stress. One mother said:

282: "It has broken my spirit & heart."

Seven mothers explained that this uncertainty has increased positive emotional health. These mothers explained that they are stronger, grateful for their child, and more able to see each day as a gift.

105: "It has made me try to enjoy every day."

Planning

The presence of uncertainty had both positive and negative effects on the ability and process of planning for the future. Nine mothers expressed that the uncertainty has prompted them to plan more for the future of their family and child, most often about specific plans such as education, long-term care, and living situations. Seventeen mothers conveyed that the uncertainty has made it more difficult and sometimes impossible to plan for the future. Most of these comments articulated the frustration and inconvenience of this inability to plan.

190: "Sometimes I feel my husband & I are paralyzed about planning for the future."

Twenty-four mothers framed this inability to plan for the future in a different way, that they take life day by day and live in the moment.

253: "Because of the uncertainty of DMBD. I'm blessed to have learned to live one day at a time. Live each day to the best of my ability."

Intrusive thoughts

Mothers frequently referred to the frequency to which they think about the uncertainty or the extent to which the uncertainty affects their life. Eighteen mothers reported thinking about the uncertainty often, and 7 explained that DBMD-related uncertainty permeates every aspect of their life. In contrast, 21 mothers described that the uncertainty does not affect them or she doesn't think about it. One mothers explained that she has normalized the uncertainty.

298: "It doesn't, really. It's just life and you have to take one day at a time and be grateful for every day."

Another mother explained that it is because her child is still quite young.

265: "Not much, he is only 8."

Practical and Financial

Sixteen mothers reported that uncertainty affects their life through practical and financial concerns. Eight participants described how uncertainty has shaped their housing situation, choice to move, and state residency. Finances and stress of saving up for medical equipment and paying medical bills was mentioned by eight mothers.

267: "I worry about being able to provide the necessities for his care. I worry about being able to afford what he needs and then worry about being able to fulfill as many dreams and have as many joyful experiences as possible in the midst of feeling the demand to save for the needs."

Mothering

Seventeen mothers explained that their child's DBMD has influenced their mothering style, approach, or capacity. One mother explained that she is overprotective, 2 explained that they spoil their child, and 3 said that they facilitate their child's social experience. Additionally, 7 said that they try to act like their son is normal and let him experience what other children do, and 2 explained how they help their child prepare for the future.

341: "I don't want to push him but I want him to experience things before he is unable. We are traveling more now, doing more fun things with him."

305: "I am often trying to help him develop friendships. If a friend calls and can get together with him, I will drop everything to make it happen."

Personal

Six mothers explained that uncertainty about their child's DBMD has influenced their faith. Five of these responses described a positive influence on their level of faith and religiosity, and one described a negative effect on faith.

369: "Given me more faith and made me search god more."

Additionally, three mothers explained that this uncertainty has taken a toll on their health, impacting sleep and general health.

450: "I wake up almost every night between 1 and 4 a.m. and can't get back to sleep, thinking about his future."

Additionally, five mothers explained career changes, pursuits, or ending that have due to their child's DBMD.

Family

Mothers also explained how uncertainty affected their families. Four mothers mentioned that their marriages have been negatively impacted, including that they get little time alone as a couple and lack social support from other couples. Additionally, five mothers explained their concern of how the uncertainty was impacting their other children. These concerns included the ability for their other children to have "normal" lives and the balance of meeting the needs of all of their children. One mother explained the extent to which uncertainty affects her family.

267: "I worry about being able to care for my other 2 boys and myself. And will my husband and be able to love and support each other through this."

Open-Ended Responses about Hope

Question 5: Many parents have described their hopefulness as changing over time. Can you describe any changes in your feelings of hope since your child was diagnosed with DBMD?

Two-hundred and nine mothers provided responses to this free response question. Responses varied in length from a word or a statement to several sentences. The statements were first coded with 1 of 5 themes of how the mothers reported their level of hopefulness changing: More hopeful/still hopeful, less hopeful, dynamic hope, shift to realistic hope, and repositioned hope. Most mothers then explained the reason for the change in their hope level. Therefore, if applicable, responses were then coded with a subcategory identifying the reported reason for the hopefulness change (Table 16). Some of the responses did not answer the prompt and were not coded.

Table 16. Quantitative Analysis of Open-Ended Question 5

How has your hope level changed? What has caused this change?	N
More Hopeful/Still Hopeful	**67**
Research/impact of future treatments	35
Son is physically doing well	10
Other	9
Faith	7
Competence/mother	6
Son is emotionally doing well	3
The positive impact of son	2
Hope for the best, prepare for the worst	2
Change in diagnosis	2
Less Hopeful	**48**
Research not fast enough	26
Son losing physical abilities	14
Other	6
Pain/Suffering	2
Repositioned Hope	**22**
Quality of Life	13
Gratitude/day-day	5
Faith	4
Dynamic Hope	**11**
Day-to-day	6
Related to son's health	5
Realistic Hope	**10**
Future treatments won't help in time	6
Realistic expectations	4

More Hopeful/Still Hopeful

Seventy-five mothers explained that they have stayed hopeful or become more hopeful over time. The most common reason that mothers cited for this change was the reality of research breakthroughs and impact of future treatments.

263: "I have become more hopeful. With treatments on the horizon, I continue to hope for a cure or at least a treatment. This is far from the devastation that I had at diagnosis."

Thirteen mothers identified that their increasing or steady hopefulness came from their child doing better physically or emotionally than they expected.

167: "Our HOPE has changes in that he is still doing so well at his age of almost 9. At early diagnosis, we were worried that he would not be doing as well as he is now. HOPE is a word that is used often in our vocabulary and we grasp on to that every chance we get."

Other common reason for the increase or stable hopefulness included faith (n=7), a 'hope for the best, prepare for the worst' mentality (n=2), and competence in mothering (n=6). One mother explained the oddity of her hope increasing, even as her child's physical abilities have decreased.

182: "I felt a lot of despair when my son was younger and the diagnosis was newer. As he has gotten older I have healed emotionally and was able to regain ground in my determination to keep living my life to the fullest. It is odd but as my son has gotten more physically needy I have felt more positive. I think this is because I feel a sense of competence in my ability to take care of him even as he needs more care."

Less Hopeful

Forty-eight mothers expressed that they have become less hopeful about their child's DBMD over time. Similar to the hopeful/still hopeful theme, the most common cited cause of mother's decrease in hope over time is that the research is not fast enough and not going to benefit her child. Other mothers reported that as their child loses physical abilities, they lose hope.

103: "Initially, we were hopeful about research, but as he gets older and we still aren't realistically closer to an answer - my hope is dwindling."

296: "It's been 4 yrs since diagnosis. I think our hope tears away just a little every time I see him lose the ability to do something."

Repositioned Hope

Twenty-two mothers explained their hope as having been shifted or repositioned. This was most commonly a shift from hope for a cure or treatment to hope for a good quality of life for the child. Several mothers cited their faith in repositioning their hope, and still others mentioned their gratitude for what they have or their son's ability to have a higher quality of life.

111: "At first, there was hope for a miracle. Today my son seems to have a slower progression and we hope that he will qualify for some of the new treatments soon that are just out of reach. So now, we have hope that he'll have a significantly less severe progression and will make it through adolescence with a great deal of independence."

340: "It becomes harder to have hope for your own visions of your child's future. You begin to realize that you have to let go and accept what comes as God's plan."

343: "Lord, yes! Before, the possibilities for his future were endless. Our goals for him were different; we pushed him to do better, always better, to run further, to jump, to be stronger…. I don't care about any of that anymore. That boy is stronger than anyone I have ever known. He never did quit, he always pushed through, and kept trying, and never stopped trusting that we were doing what we

thought was best for him. I know he'll never be the fireman or drummer in a rock band that we hoped for when he was younger, and discovering life, finding the things he loved, but that's okay. I have buried that future. He may not save lives by running into burning buildings, but he has already been inspirational for many people, and given hope that if he can keep getting up and climbing his mountain, the rest of us can, too. My only hope [for] him now, is for a cure, or treatment that will allow him to live a good, long life, and to experience all the things he wants to, how he wants to."

Dynamic Hope

Another theme reported by mothers is that their level of hope has gone up and down over time, depending on the circumstances. The most common reason for this fluctuating level of hope included the health status of the child and treatment changes, but mothers also explained that these hope shifts occur on a daily basis due to many circumstances.

305: "I try to not to look too far ahead and make plans. It's my way of guarding myself because it does change. One day I can feel very encouraged, and then I the next I can get a discouragement in either an event, or a call and that hope can change both for good or not so good."

426: "First month completely hopeless, then roller coaster of emotions, drug to be approved then not then approved etc.....everything takes way tooooo long!"

390: "My feelings of hope are constantly changing. I feel completely hopeless that a cure will be found or that he will ever get better, but I remain hopeful that he will live a fulfilling life."

Realistic Hope

Ten mothers explained their hope as being realistic. Several articulated that although they do have hope, they still are able to be realistic about what the expectations are for their son and their son's future.

281: "I try to be as realistic as I can, not that I don't have hope, just that I know what the future holds and I try not to get lost with other expectations."

119: "We continue to hope for a cure but the reality of the situation is so uncertain that we know a cure will not benefit him."

DISCUSSION

Uncertainty

The mothers in this study reported relatively low uncertainty on the PUCHS uncertainty scale, as indicated by a mean weighted uncertainty score of -0.71 on a -2 to 2 scale, with more negative values indicating less uncertainty. These mothers of children with DBMD had less uncertainty than parents of children with chronic undiagnosed conditions (Macnamara et al., 2014). While parents must face prognostic and medical management uncertainties after DBMD diagnoses, parents without a diagnosis for their child face not only more prognostic and medical management uncertainty but also diagnostic uncertainty. In our study, the dimension with the highest uncertainty for mothers was medical management, which was affirmed by the 58% of mothers who qualitatively articulated that their child's prognosis, lifespan, and medical care plan was the most uncertain to them. Although neither social uncertainty or reproductive uncertainty were reported by any participants as most uncertain in the free response section, the PUCHS data revealed that they do quantitatively perceive some uncertainty within these dimensions. Mothers had the lowest level of uncertainty within the existential dimension. The existential uncertainty questions on the PUCHS scale are quite broad and address the mother's perspective of her child's life as a whole, i.e. "My child's diagnosis of DBMD leaves me uncertain about the meaning or purpose of my child's life". Although mothers did not perceive general existential uncertainty about their child's life, as reported by the PUCHS, the qualitative responses that came closest to expressing existential uncertainty addressed uncertainty about the child's life experiences specifically and how to best use the time that their child has. Thirty-eight percent of

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mothers mentioned these child's life experience uncertainties, which include uncertainty about their child's quality of life, educational and career opportunities, social life, and future independence. In other words, mothers may have more specific existential uncertainties but not perceive the purpose of their child's life as uncertain. If the existential questions on the PUCHS were more specific, the uncertainty level of this dimension may be higher.

Each uncertainty dimension was predicted by a different array of child and mother characteristics, demonstrating that these dimensions of uncertainty are distinct and impacted by different factors. Participation in a clinical trial was associated with less medical management and social uncertainty. This association could be due to social support present within this community, a positive outcome from the trial, or possible access to a higher level of expertise and care through the specialty centers acting as clinical trial sites. Mothers with younger children perceived more medical management uncertainty, reproductive uncertainty, and social uncertainty. This decrease in these various uncertainties as children get older could be attributed to mothers having more experience with medical management and being able to envision a trajectory for their child compared to other children with DBMD. Social uncertainty might recede as mothers become more comfortable in their new role as parents of a child with DBMD and build relationships the offer social support. As expected, reproductive uncertainty was significantly associated with child's age, mother's age, and carrier status; younger mothers who are carriers and have younger children are likely still making reproductive decisions and are at risk for having another child with DBMD. The number of children affected was associated with medical management uncertainty and existential uncertainty.

It seems that already having a child with DBMD does not provide a sense of assurance in being experienced; having more children with DBMD actually amplifies the perceptions of medical and quality of life-related uncertainty. This could be attributed to the added complexities and stress of financially, physically, and emotionally caring for more than one child with DBMD. Multivariate analysis revealed that mother's age is a statistically significant independent variable of total uncertainty after controlling for hope and child's functional status, while bivariate analysis revealed that maternal age is only significantly associated with medical management and reproductive uncertainty. Combined, the two analyses suggest that uncertainties within these two dimensions are driving the relationship seen between total uncertainty and mother's age.

Relationships between Uncertainty and Predictor Variables

We hypothesized that there would be a relationship between hope and uncertainty, as qualitative analyses have explained that hope's role is to provide "possibilities within uncertainty" (Duggleby et al., 2010). In multiple linear regression, increased hope, decreased child's ambulation, and increased maternal age were all independently associated with less uncertainty. This finding is supported by a study of cancer survivors where hope was significantly correlated with lower levels of perceived uncertainty (Wonghongkul et al., 2000). Although this relationship is plausible, the cross-sectional results presented here do not inform the directionality of the relationship, and this study cannot determine whether having higher hope results in lower uncertainty or lower uncertainty causes increased hope.

We also hypothesized that mothers of children with less ambulation would have greater uncertainty, because as a child loses functionality, they become closer to death

and the uncertainty when death may occur becomes more evident. Data refuted this hypothesis, indicating that mothers of children with more progressive DBMD perceived less uncertainty. Mishel's Uncertainty Theory asserts that uncertainty surrounding an illness typically decreases with age for most chronic or stable conditions (Mishel, 1988); our data suggests that this trend is also true for progressive conditions. This finding could be due to several factors. First, it is possible that mothers of children with less ambulatory abilities perceived less uncertainty because there is less to be uncertain about. This possibility is supported qualitatively in this study, where 21% of mothers reported feeling most uncertainty about their child's physical prognosis and when they will lose certain abilities. A second possibility is that as their child's condition progresses, mothers have had more time to process the diagnosis and its associated uncertainties and may therefore be appraising the uncertainty differently, resulting in less perceptions of uncertainty. This third hypothesis is supported qualitatively; several mothers explained that the period immediately following diagnosis was filled with anxiety, fear, and uncertainty, but over time, as they have learned more about DBMD, found a trustworthy medical team, and met other DBMD families, some of these negative psychological emotions and uncertainty have either subsided or been replaced.

Mother's age had a statistically significant negative relationship with uncertainty, where younger mothers reported more uncertainty. Although increasing mother's age is related to a decline in child's abilities, the association between mother's age and uncertainty must be attributed to reasons beyond those suggested for the relationship between uncertainty and functional status, because the relationship between mother's age and uncertainty is independent of functional status. A similar association between

maternal age and uncertainty was seen in a study of parents of children with rare chromosome conditions (Lipinski et al., 2006), but the opposite relationship was found in a study of parents who have children with Down syndrome (Truitt et al., 2012). Greater levels of uncertainty in younger mothers could stem from the limited experience that younger parents have with the diagnosis and healthcare systems. Further, the relationship between maternal age and uncertainty may also be explained by the fact that older individuals have fewer reproductive plans, and therefore less reproductive uncertainty.

We also hypothesized that mothers of children with Becker muscular dystrophy would have greater uncertainty because the presentation of Becker is more variable and could result in more prognostic uncertainty. Although this relationship did not prove to be statistically significant, three mothers qualitatively reported that the variability of progression in BMD, in contrast to the more predicted progression of DMD is what incites the most uncertainty for them. The lack of a statistically significant relationship could be attributed to the lack of power and small sample size of mothers of a child with BMD in our study (11%).

Relationships between Coping Efficacy and Predictor Variables

Coping efficacy is an appraisal of how confident a parent feels about their ability to cope with their child's DBMD medical condition. In our conceptual model, coping efficacy is an appraisal that affects coping and ultimately adaptation. Better understanding of the factors that predict higher coping efficacy may lead to the development of interventions designed to facilitate parental adaptation to uncertainty. In multivariate analysis, uncertainty, hope, and spirituality remained statistically significant predictors of coping efficacy.

Hope was found to have a positive relationship with coping efficacy; mothers with greater levels of hope view themselves as better able to cope with their child's DBMD. These results affirm conclusions from other studies finding hope to be positively associated with similar outcome variables including well-being, quality of life, and adaptation (Bailey et al., 2007; Davis, 2005; Truitt et al., 2012). Optimism, faith, and wishful thinking are all related constructs to hope. Hope is influenced by both internal and external factors, with one of the internal factors being spirituality. Interestingly, we found that spiritualty had a statistically significant relationship with coping efficacy independent of the role of hope, affirming that our hope scale is measuring a separate concept from spirituality.

In this population, mothers who had higher uncertainty had lower coping efficacy. Mishel's Uncertainty in Illness Theory states that uncertainty can be appraised as either a threat or an opportunity, that parents can be either burdened by or grateful for the uncertainty that exists as part of their child's diagnosis; how mothers choose to appraise this uncertainty will impact their adaptation (1988). Our results suggest that these mothers appraise this uncertainty as both a threat and an opportunity within a variety of different categories.

Moderation Analysis

Identifying variables that impact this uncertainty appraisal process may shape interventions for supporting mother's adaptation to uncertainty. Hope has been described as a promising variable impacting the appraisals of stressors, such as uncertainty (Miller 2007). Although the exact role of hope in adaptation to uncertainty is not understood, hope could moderate the relationship between the stressor and the adaptation process

(Folkman and Greer, 2000; Valle et al., 2006; Horton and Wallander, 2001). Therefore, we hypothesized that hope functions as a moderator causing the relationship between uncertainty and coping efficacy to vary by the level of hope. The moderation analysis in this study revealed that such an interaction between hope and uncertainty was not significant. Our results parallel those of a study looking at the role of hope in the relationship between uncertainty and adaptation in caregivers of children with Down syndrome (Truitt et al., 2012), although hope has been seen as a significant moderator in relationships of key variables of similar concepts (Valle et al., 2006; Horton and Wallander, 2001). There could be several reasons why this interaction did not reach significance in our study. First, the level of uncertainty reported in this population was quite low, and it is possible that in a population with greater variance in uncertainty hope may emerge as a significant moderator. Additionally, it is possible that hope moderates the relationship between uncertainty and a different outcome, such as quality of life, or life satisfaction, or does not serve as a factor in the uncertainty appraisal process.

Hope

The results of free response question about hope allow us to better understand the concept of hope and its role in adapting to uncertainty. Previous hope scales have operationalized hope as a trait variable (Herth 1989). Others have sought to define hope as a state-dependent variable, changing over time and understood within the context of uncertainty, but this concept has not been well operationalized (Dufault and Martocchio, 1985). The novel state hope measure used in this study was shaped by existing interview studies of parents of children with DBMD but needed additional content validation (Duggleby et al., 2010; Samson et al., 2009). The majority of mothers in this study

qualitatively affirmed that they viewed their hope as changing over time, and most cited a reason for the change. Interestingly, most mothers cited research and the existence of new treatments on the horizon as a reason for hope changes in both the more hopeful and less hopeful direction. This qualitative dichotomization of the way hope levels can shift exemplifies why hope may be a factor in uncertainty appraisals. These mothers all share the stressor of uncertainty of their child's prognosis. Some view the uncertainty of possible treatments down the road as hope-instilling (appraising uncertainty as an opportunity) whereas other mothers view this same uncertainty as hopeless and unrealistic (appraising uncertainty as a threat). Hope might not have been a statistically significant moderator of the uncertainty and coping efficacy relationship, but hope did have an independent effect on uncertainty and coping efficacy. Therefore, both the qualitative and quantitative data uphold hope as a considerable factor in this appraisal process. Some of these mothers also explained that they have since repositioned their hope, often from the hope of a cure to the hope of a high quality of life for their child. This explanation of the repositioning of hope continues to support our conceptualization of hope as a dynamic situation-dependent variable.

Clinical Implications

Findings from this study have implications for healthcare providers, including genetic counselors, who work with families of children with chronic progressive medical conditions such as DBMD. This study contributes to the understanding of factors that impact uncertainty appraisals, coping efficacy, and ultimately adaptation to a child's condition. Younger mothers, lower hope scores, and having ambulatory children were associated with more uncertainty. Additionally, lower hope, higher uncertainty, and less

reported spirituality were associated with lower coping efficacy. Although the directionality of these relationships cannot be evaluated from this study, the Uncertainty in Illness theory and the Transactional Model of Stress and Coping postulate that uncertainty is an antecedent of coping efficacy, which is an antecedent of adaptation. Therefore, techniques aimed at reducing or reframing uncertainty could increase coping efficacy, as well as adaptation. Although it may not be possible to mitigate uncertainty, especially within the medical management and prognostic uncertainty domains, efforts to help mothers manage uncertainty may be more effective if tailored towards specific uncertainties that are more salient to them. For example, because younger mothers with more ambulatory children report more uncertainty, offering extra support and education to families with a newly diagnosed child may help alleviate the initial uncertainty of the condition and diagnosis.

Because hope is negatively associated with uncertainty and positively associated with coping efficacy, hope seems to be a factor in shaping uncertainty appraisals, facilitating coping efficacy, and possibly promoting adaptation. Since we operationalized hope as a disease-focused state and not a trait factor, it may be possible for healthcare providers such as genetics counselors to bolster hope in low-hope individuals. Many of the mothers in this study explained that research and the hope for future treatments elevate their hope levels, but other mothers explained the exact opposite, that the lack or slow development of treatments had had a negative impact on hope levels. Some of these mothers explained that they have since repositioned their hope often from the hope of a cure to the hope of a high quality of life for their child. Although more research is needed to develop specific hope interventions, this data suggests that a possible hope

instilling intervention could assist mothers in redirecting hope even if it is lost in another area. Even if hope is not easily fostered, guiding mothers with low hope to other uncertainty management and reappraisal strategies may be helpful.

Limitations

While this data has clinical implications for healthcare providers, there are several limitations to the study. Due to the cross-sectional nature of this study, assessments can only be made about how the variables are associated with one another, rather than being able to determine the direction of the relationships.

Another potential limitation is the possibility of recruitment bias. Although the representativeness of the study sample was increased by the diversity of recruitment sources, participation bias may have been generated due to the opt-in nature of this study. For example, it is possible that mothers of children with DBMD who chose to participate in this study have more or less hope or uncertainty than those who chose not to participate. Additionally, because one of the recruitment sources is a registry, is it possible that these mothers and their children have been more involved with research, clinical trials, and support resources. Recruitment was also done at two time points, with the recruitment of the second time point being primarily from the Duchenne Connect registry approximately 3 years after the original recruitment. Comparison of these two recruitment groups indicated that the second recruitment group had statistically significant differences in the child's current age (Δ=2.8 years), mother's age (Δ=3.8 years), child's functional status, and uncertainty level. Since these groups were recruited about 3 years apart, it makes sense that the second recruitment group would be younger, have younger and more functional children, and perceive more uncertainty (because

uncertainty is negatively related to child's functional abilities). It is also possible that these groups differed by variables that were not assessed by this study. Additionally, the originally recruited participants completed 2 similar surveys in the longitudinal study prior to this survey and therefore had exposure to similar scales and may have answered differently due to this experience.

The participant characteristics introduce another potential limitation in this study. The study population was largely non-Hispanic, Caucasian, married, and well-educated, and thus the results of this study may not be generalizable to the greater population of mothers of children with DBMD. Additionally, several mothers of adult children with DBMD contacted us and expressed that some of the items on the uncertainty and hope scales were difficult to answer and not applicable to their circumstances of caring for their adult child. Mothers were allowed to skip questions that did not pertain to their situation. About 25% of the mothers in this study had children over the age of 18. Therefore, we are designing a follow-up survey tailored to mothers who have adult children.

Areas for future research

The plans for future analysis of this data include completing further qualitative analysis of the open-ended questions addressing what mothers find most uncertain, how they are appraising this uncertainty, and how they feel that their hope levels have changed over time. Once thematic analysis and second coding is complete, we aim to see whether hope levels impact qualitative uncertainty appraisals, therefore potentially identifying hope as a moderator in the relationship between uncertainty and coping efficacy.

Hope has primarily been operationalized as a trait variable and not a state, situational-dependent variable. This is the first study to use this novel hope scale, so further validation of this measure is needed. Additionally, the impact of increasing state hope on long-term adaptation is an area for future study, as well as developing an evidence-base for hope bolstering and repositioning interventions.

Prospective data are needed to make casual inferences, and since this study is imbedded within a longitudinal study, we hope to include the uncertainty, hope, and coping efficacy measures in future surveys to gain insight into the directionality of these relationships and how they change over time.

Conclusion

This study of mothers of children with DBMD identified important relationships between uncertainty, hope, and coping efficacy. Because younger mothers of more ambulatory children with DBMD perceive more uncertainty, especially uncertainty related to medical management and social support, efforts to help mothers manage uncertainty may be more effective if tailored towards mothers of children with new diagnoses and specific domains of uncertainties most salient to them. Additionally, hope seems to be a factor in shaping uncertainty appraisals and facilitating coping efficacy. Although future studies are needed, interventions aimed at bolstering maternal hope or guiding mothers with low hope to other uncertainty management and reappraisal strategies may be helpful.

APPENDIX A: Pre-Survey Announcement

Hello and happy summer! Thank you for your participation in our project on the wellbeing of mothers of children with Duchenne or Becker muscular dystrophy. We hope that this message finds you and your family well. We've been busy analyzing the data from this study and have two articles we'll submit before the end of the summer. We've also presented data on this study in front of hundreds of clinicians and researchers and have used the data to support two grant applications—and we're just getting started. We'll send you a summary of the findings to date later this year.

You will receive your next survey in about 2-3 weeks. Because we are half way through this study, it's time for a small thank you—at the end of the survey you'll receive a $20 Amazon.com gift card.

Have any events happened since last survey that had a profound effect on your life or your family life that we should know about for our study? If so, please respond to this message to give us a brief update.

In the sad event that your child with DBMD has died since you completed your last survey, please let us know by sending a brief response to this email.

Look for an email from us in the next 2-3 weeks with your survey link! Until then, enjoy the sun.

Best,
Holly Peay
PPMD
holly@parentprojectmd.org

Study Contact Information:
For questions, concerns, or complaints about this research study or to report a research-related injury, or for information about research participants' rights, you can contact the researcher Holly Peay, MS CGC at Parent Project Muscular Dystrophy at 443-791-5927. Researchers are available to answer any questions you may have about the research at any time.

APPENDIX B: Study Advertisement for Duchenne Connect and Clinicaltrials.gov

Wellbeing in Mothers Caring for a Child with Duchenne or Becker Muscular Dystrophy

The National Human Genome Research Institute within the National Institutes of Health and Parent Project Muscular Dystrophy are conducing a research study to learn more about the experiences of mothers caring for a child with Duchenne or Becker muscular dystrophy (DBMD).

Study goal

This study will help us to determine how the experiences and wellbeing of mothers caring for a child with DBMD change over time. The overall goal of the study is to obtain information to support the development of interventions for mothers that aim to improve their wellbeing.

Who can participate?

This study is open to English-speaking females, 18 years of age or older, who are biological mothers of one or more children with DBMD. The child with DBMD can be any age. All participants must currently be residing in the U.S. and be willing to participate in this study for 3 years. If you have previously joined this study, please disregard this advertisement.

What is involved in participating in the study?

Participation involves agreeing to be a part of this study and answering the surveys that will be sent to you either online or in the mail. After the initial survey has been completed, shorter follow-up surveys will be sent approximately every 6-12 months for about 3 years. You will be a part of this study for approximately 3 years unless you choose to withdraw.

What are the risks of the study?

The potential risks for participating in this study are minimal, but may involve becoming emotionally upset because of the questions asked in the survey. For that reason, participants will be provided with the contact information for a genetic counselor and support group.

When is this study taking place?

Recruitment for this study will be ending on October 31st, 2015.

For more information, contact study investigator Holly Peay at holly@parentprojectmd.org or 443-791-5927 or Megan Bell at bellme@mail.nih.gov or 605-261-5927.

APPENDIX C: Consent document for new participants, Posted at the Beginning of the Survey

Assessing Wellbeing in Women Caring for Children with Duchenne or Becker Muscular Dystrophy

INVESTIGATOR INFORMATION:
Holly Peay, MS CGC Parent Project Muscular Dystrophy, Associate Investigator
Megan Bell, National Human Genome Research Institute, NIH, and Johns Hopkins
      School of Public Health, Associate Investigator
Martha E. Walker, MS, CGC, Division of Human Genetics, Principal Investigator

INTRODUCTION:
You are being asked to take part in this research study because you are the birth mother of a child(ren) who has been diagnosed with Duchenne or Becker muscular dystrophy (BDMD). It is your choice whether to participate in this research study.
This study proposes to examine the concept of wellbeing in women caring for children with Duchenne/Becker muscular dystrophy (BDMD) in order to develop interventions designed to improve caregiver's quality of life. The study aims to describe perceived needs and motivations of mothers of children with DBMD and explore a range of predictors of wellbeing. The long-term outcome of this study is to inform a set of accessible interventions to help women achieve a greater degree of overall wellness.

To participate in the study, you must:
1) Be a birth parent of a child with Duchenne or Becker
2) Live in the United States
3) Be able to answer a survey in English
4) Be at least 18 years old

Your child with DBMD can be any age.

This study will last for about 3 and a half years, with surveys every 6-12 months. The surveys will take about 30-45 minutes to complete. The surveys will include information about your needs as a mother, information about your own thoughts and feelings, and measures such as self-concept and adaptation. You can answer the surveys online or you can receive paper copies of the surveys.

You may withdraw from the study at any time without penalty. If you decide not to participate, there will be no penalty or loss of benefits to which you are otherwise entitled.

WHAT ARE THE RISKS AND BENEFITS OF THE RESEARCH STUDY?
This is considered a minimal risk study. On rare occasion, participants may experience emotional distress and/or discomfort from discussing topics related to being a mother of a child with DBMD. If needed, genetic counselors will be available to speak with patients and/or parents.

If you agree to take part in this research study, you will not receive a direct medical benefit. The significance of the study will be learning new information to be applied to the development of a well being intervention program for mothers of children with DBMD.

WHAT WILL YOU GET FROM JOINING THIS STUDY?

You will receive a $20 Amazon gift card for completing the first survey. An electronic gift card will be emailed to you following completion of the survey unless you request a gift card be mailed to you.

HOW WILL INFORMATION ABOUT YOU BE KEPT PRIVATE AND CONFIDENTIAL?

You will be assigned a study identification code so that your study information will be confidential. The link between your name and study identification code will be kept by the study research staff in a secure environment. The results of the study may be published, but your name or identity will not be revealed. To ensure that confidentiality is maintained, the principal investigator and research staff will keep records in secure offices. Your research record will be available only to study staff.

WHO DO YOU CALL IF YOU HAVE QUESTIONS OR PROBLEMS?

For questions, concerns, or complaints about this research study or to report a research-related injury, or for information about research participants' rights, you can contact the researchers

Holly Peay, MS CGC at Parent Project Muscular Dystrophy at (443) 791-5927 or Megan Bell at (605) 261-5927. Researchers are available to answer any questions you may have about the research at any time.

AGREEMENT TO PARTICIPATE IN SURVEY:

I have read the information given above and I agree to participate in this survey. ☐

APPENDIX D: Full Survey including measures for the longitudinal study

Thank you for participating in this study about the feelings, thoughts, and needs of mothers caring for children with Duchenne or Becker muscular dystrophy (DBMD). This is the 3rd survey. You will receive follow up surveys about every 12 months for the next 3 years. This survey may take you about 30-45 minutes to complete. You do not have to finish all of the survey at one time, but we encourage you to answer all of the questions. There are no correct or incorrect answers.

While we use the term "child" in this survey, we understand that some participants have children who are teens or adults. We are interested in the thoughts and feelings of mothers of adult children with DBMD as well as mothers of younger children.
We appreciate you sharing your thoughts and experiences so we can create interventions that will help mothers' wellbeing.

WHO DO YOU CALL IF YOU HAVE QUESTIONS OR PROBLEMS?

For questions, concerns, or complaints about this research study or to report a research-related injury, or for information about research participants' rights, you can contact Holly Peay at Parent Project Muscular Dystrophy at 443-791-5927 or holly@parentprojectmd.org. Please also contact Holly Peay if you would like another copy of the consent statement.

** Please follow the instructions at the beginning of each section. Thank you for your time and participation.**

SECTION A: Questions about Your Children (For Returning Participants)

Update on Your Children
1. Have you given birth to any children in the past 12 months?
   a. Yes
   b. No

2. [If yes] What is the sex of the baby? M/F
3. Does this child have Duchenne or Becker? Y/N/Unsure

4. Have any of your children died in the past 12 months?
   a. Yes
   b. No

5. [If yes] Did this child have DBMD? [If yes, send them to the alternate survey]

About your child with DMBD
1. Chose the option that best describes your child's physical abilities today. If you have more than one child with DBMD, please answer this question about your **oldest** child. Every child is unique, and may not match the descriptions perfectly. Please select the answer that is **the best fit**. My child with DBMD:
   a. Presymptomatic – Has no symptoms
   b. Early-ambulatory – Walks with an unusual gait but is able to climb stairs
   c. Late-ambulatory - Walks with more difficulty, sometimes uses a wheelchair, is losing the ability to get up from the floor and climb stairs
   d. Early non-ambulatory – Is unable to walk alone but can still sit and stand, uses a non-powered wheelchair on his or her own
   e. Non-ambulatory I- Uses a powered wheelchair but is no longer able to use a non-powered wheelchair on his or her own; is showing limited arm strength though is able to raise hands to mouth
   f. Non-ambulatory II– Is no longer able to raise hands to mouth but is able to hold a pen or to move powered wheelchair
   g. Non-ambulatory III- Is no longer able to use his/her hands to hold a pen

2. Has your child ever participated in a DMBD related clinical trial?
   a. Yes
   b. No

SECTION A: Questions about Your Children (For New Participants)

About your children

1. Please describe your children, from OLDEST to YOUNGEST

	Year of birth	Sex M/F	Does this child have a neuromuscular disorder? Yes (list diagnosis), No, Don't know	Is this your biological or adopted child?	Is this child still living? Y/N
Child 1					
Child 2					
Child 3					
Child 4					
Child 5					
Child 6					
Child 7					
Child 8					
Child 9					

About your child with DMBD

1. At what age was your child with DBMD diagnosed? (If you have more than one child with DBMD, please answer for the oldest child.)
    a. Prenatally
    b. 0-3 years
    c. 4-7 years
    d. 8-11 years
    e. 12 years or older

2. Before your child was diagnosed, did you know any blood relatives with DBMD? Y/N

3. Chose the option that best describes your child's physical abilities today. If you have more than one child with DBMD, please answer this question about your **oldest** child. Every child is unique, and may not match the descriptions perfectly. Please select the answer that is **the best fit**. My child with DBMD:
    a. Presymptomatic – Has no symptoms
    b. Early-ambulatory – Walks with an unusual gait but is able to climb stairs
    c. Late-ambulatory - Walks with more difficulty, sometimes uses a wheelchair, is losing the ability to get up from the floor and climb stairs
    d. Early non-ambulatory – Is unable to walk alone but can still sit and stand, uses a non-powered wheelchair on his or her own
    e. Non-ambulatory I- Uses a powered wheelchair but is no longer able to use a non-powered wheelchair on his or her own; is showing limited arm strength though is able to raise hands to mouth
    f. Non-ambulatory II– Is no longer able to raise hands to mouth but is able to hold a pen or to move powered wheelchair
    g. Non-ambulatory III- Is no longer able to use his/her hands to hold a pen

4. Has your child ever participated in a DMBD related clinical trial?   Yes/No

SECTION B: Your Resilience

This section includes questions about how you feel about yourself. Please answer the following questions by choosing the option that shows how much you believe each statement is true.

	Strongly Disagree	Disagree	Neutral	Agree	Strongly Agree
I enjoy being with other people	1	2	3	4	5
It is easy to be flexible in social situations	1	2	3	4	5
I have friends/family members who appreciate my abilities.	1	2	3	4	5
When I have a goal, I do my best to attain it.	1	2	3	4	5
I establish friendly relationships easily.	1	2	3	4	5
I enjoy being with my family.	1	2	3	4	5
I communicate well with new people.	1	2	3	4	5
When in difficult situations, I know there is a better future	1	2	3	4	5
There are strong connections among my friends.	1	2	3	4	5
I laugh easily.	1	2	3	4	5
I can discuss personal issues with friends/family members.	1	2	3	4	5
I believe in my abilities.	1	2	3	4	5
There are family members/friends who help me.	1	2	3	4	5
I know how to achieve my goals.	1	2	3	4	5
My family agrees on important affairs in life.	1	2	3	4	5
I can solve my personal problems.	1	2	3	4	5
Regular rules make my daily life easier.	1	2	3	4	5

It is easy to find subjects to talk about with other people.	1	2	3	4	5
I know I will succeed if I keep trying.	1	2	3	4	5
I have friends/family members who encourage me.	1	2	3	4	5
I prefer to have plans for my activities.	1	2	3	4	5
My family is optimistic in difficult situations.	1	2	3	4	5
I trust my judgments and decisions.	1	2	3	4	5
There is always someone who helps me when needed.	1	2	3	4	5
I am quickly informed when a family member has a problem.	1	2	3	4	5
I have strong connections in my family.	1	2	3	4	5
A good future awaits me.	1	2	3	4	5
My family is honest with each other.	1	2	3	4	5
I maintain daily rules even in difficult situations.	1	2	3	4	5
My family enjoys finding a chance to do things together.	1	2	3	4	5
I always find a way to solve problems regardless of what happens.	1	2	3	4	5
I have realistic plans for the future.	1	2	3	4	5

SECTION C: Your Coping with DBMD

Instructions: Many mothers use different ways to cope with caring for a child with DBMD. This section asks you to rate how well you can perform different types of coping. Choose the number (1-11) that best shows to how confident or certain you are that you can do what is described to cope with your child's DBMD.

Cannot do at all				Moderately certain can do						Certain can do
1	2	3	4	5	6	7	8	9	10	11
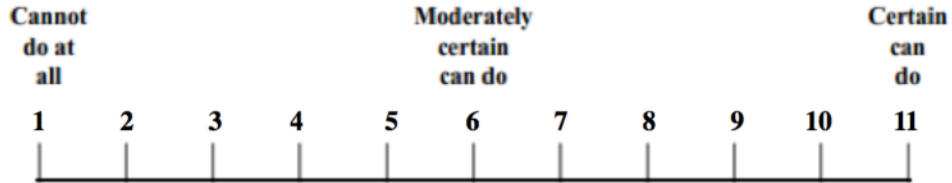
When things aren't going well for you, or when you're having problems, how confident or certain are you that you can:

1. Break an upsetting problem about DBMD down into smaller parts _____
2. Sort out what can be changed and what cannot be changed about DBMD _____
3. Make a plan of action and follow it when confronted with a problem related to DBMD _____
4. Leave options open when things related to DBMD get stressful _____
5. Think about one part of a DBMD problem at a time _____
6. Find solutions to your most difficult DBMD problems _____
7. Resist the impulse to act hastily when under pressure _____
8. Try other solutions to DBMD problems if your first solutions don't work _____
9. Talk positively to yourself _____
10. Stand your ground and fight for what you want _____
11. See things from another person's point of view during a heated argument about DBMD _____
12. Develop new hobbies or recreations _____
13. Make unpleasant thoughts about DBMD go away _____
14. Take your mind off unpleasant thoughts about DBMD _____
15. Stop yourself from being upset by unpleasant thoughts about DBMD _____
16. Keep from feeling sad about DBMD _____
17. Keep from getting down in the dumps about DBMD _____
18. Look for something good in a negative situation _____
19. Keep yourself from feeling lonely _____
20. Visualize a pleasant activity or place _____
21. Pray or meditate _____
22. Get friends to help you with the things you need _____
23. Get emotional support from friends and family _____
24. Make new friends _____
25. Do something positive for yourself when you are feeling discouraged _____
26. Get emotional support from community organizations or resources _____

SECTION D: Your DBMD hopes

<u>Instructions</u>:

Below are statements related to hopes for your child. How often in the **past 4 weeks** you found hope for the future in each statement? Read and choose an answer for all the statements, even if you are not completely sure about your answer.

This is what the numbers mean:
1. *Never*
2. *Almost never*
3. *Sometimes*
4. *Often*
5. *Always*

We think of "hope" as the best possibilities for your child's future. In the **past 4 weeks**, how often have you found hope in…

	Never	Almost never	Sometimes	Often	Always
Day-to-day life with your child	1	2	3	4	5
Parenting your child	1	2	3	4	5
Being able to care for your child	1	2	3	4	5
Understanding your child	1	2	3	4	5
Research being done today	1	2	3	4	5
Aspects of your child's clinical care	1	2	3	4	5
Imagining your child's bright future	1	2	3	4	5
Your confidence about your child's future	1	2	3	4	5
The uncertain potential of your child	1	2	3	4	5
Your child's long-term health could be better than expected	1	2	3	4	5

In the **past 4 weeks**, how often have you found hope, *even*…

	Never	Almost never	Sometimes	Often	Always
Given your child's diagnosis	1	2	3	4	5
When you are realistic	1	2	3	4	5

In the **past 4 weeks**, how often have you found hope *through*:

	Never	Almost never	Sometimes	Often	Always
Your relationships with others	1	2	3	4	5
Your determination	1	2	3	4	5
Your faith or spirituality	1	2	3	4	5
Your use of the resources available to you	1	2	3	4	5

SECTION E: Spirituality

The list that follows includes items which you may or may not experience, please consider how often you directly have this experience, and try to disregard whether you feel you should or should not have these experiences. A number of items use the word God. If this word is not a comfortable one for you, please substitute another idea that calls to mind the divine or holy for you.

	Many times a day	Every-day	Most days	Some days	Once in a while	Never or almost never
I feel God's presence	1	2	3	4	5	6
I experience a connection with life	1	2	3	4	5	6
During worship, or at other times when connecting with God, I feel joy which lifts me out of my daily concerns	1	2	3	4	5	6
I find strength in my religion or spirituality	1	2	3	4	5	6
I find comfort in my religion or spirituality	1	2	3	4	5	6
I feel deep inner peace or harmony	1	2	3	4	5	6
I ask for God's help in the midst of daily activities	1	2	3	4	5	6
I feel guided by God in the midst of daily activities	1	2	3	4	5	6
I feel God's love for me, directly	1	2	3	4	5	6
I feel God's love for me, through others	1	2	3	4	5	6
I am spiritually touched by the beauty of creation	1	2	3	4	5	6
I feel thankful for my blessings	1	2	3	4	5	6
I feel a selfless caring for others	1	2	3	4	5	6
I accept others even when they do things that I think are wrong	1	2	3	4	5	6
I desire to be closer to God or in union with the divine	1	2	3	4	5	6

	Not Close at All	Somewhat Close	Very Close	As Close as Possible
In general, how close do you feel to God?	1	2	3	4

SECTION F: Experience with your child with DBMD

Instructions: Please rank the degree to which you agree with the following statements from 1-strongly disagree to 5-strongly agree.
If you have multiple children with DBMD, please focus on your experience with your oldest affected child while answering the following questions.

My child's diagnosis of DBMD leaves me…

	Strongly Disagree	Disagree	Neither Agree or Disagree	Agree	Strongly Agree
Unsure how to think about my child's condition	1	2	3	4	5
With no clear understanding of my child's limitations	1	2	3	4	5
Unclear how to participate in the long-term treatment decisions for my child	1	2	3	4	5
Unsure where to go for treatment of my child's condition	1	2	3	4	5
Unsure of my child's expected lifespan	1	2	3	4	5
Unsure about having more children	1	2	3	4	5
Uncertain what to tell relatives about risks to their children	1	2	3	4	5
Unclear how to make decisions for my family not knowing what the future may hold for my child	1	2	3	4	5
Uncertain how to address my family's	1	2	3	4	5

concerns about my child					
Not knowing how to find parents in a similar situation	1	2	3	4	5
Unsure about finding support from parents going through similar experiences	1	2	3	4	5
Uncertain about the meaning of my child's life	1	2	3	4	5
Unclear about the purpose of my child's life	1	2	3	4	5

Please rank how important each item is to you from: 1-Not important to 5-Very important

	Not Important	Somewhat Not Important	Neither Important or Not Important	Somewhat Important	Very Important
Knowing how to think about my child's condition	1	2	3	4	5
Having a clear understanding of my child's limitations	1	2	3	4	5
Participating in long-term treatment decisions for my child	1	2	3	4	5
Knowing where to go for treatment of my child's condition	1	2	3	4	5
Knowing my child's expected	1	2	3	4	5

lifespan					
Having information to make decisions about having more children	1	2	3	4	5
Being able to tell relatives about risks to their children	1	2	3	4	5
Being able to make decisions for my family not knowing what the future may hold for my child	1	2	3	4	5
Addressing my family's concerns about my child	1	2	3	4	5
Finding parents in a similar situation	1	2	3	4	5
Having support from parents going through similar experiences	1	2	3	4	5
Having clarity about the meaning of my child's life	1	2	3	4	5
Understanding the purpose of my child's life	1	2	3	4	5

SECTION G: Open-ended questions
Tell us about the uncertainty you feel about DBMD.

1. Many parents explain that they feel uncertainty surrounding their child's DBMD. What is one thing about your child's DBMD that feels the most uncertain?

2. How has this uncertainty affected your life?

3. Share an example of how this uncertainty has a negative impact on you.

4. Share an example of how this uncertainty has a positive impact on you.

5. Many parents have described their hopefulness as changing over time. Can you describe any changes in your feelings of hope since your child was diagnosed with DBMD?

SECTION H: Questions about you

Instructions: The following section asks questions about your personality. Please follow the directions for each set of questions. Please answer each item.

	Strongly Disagree	Disagree	Neutral	Agree	Strongly Agree
Conflicting expert opinions about a medical test or treatment would lower my trust in the experts.	1	2	3	4	5
I would not have confidence in a medical test or treatment if experts had conflicting opinions about it.	1	2	3	4	5
Conflicting expert opinions about a medical test or treatment would make me upset.	1	2	3	4	5
I would not be afraid of trying a medical test or treatment even if experts had conflicting opinions about them.	1	2	3	4	5
If experts had conflicting opinions about a medical test or treatment, I would still be willing to learn them.	1	2	3	4	5
I would avoid making a decision about a medical test or treatment if experts had conflicting opinions about it.	1	2	3	4	5

SECTION I: Burden

The questions below reflect how people sometimes feel about **their child with DBMD**. After each statement, please circle the response that best describes how often you feel this way. There are no right or wrong answers.

Do You Feel:	Never	Rarely	Sometimes	Quite Frequently	Nearly Always
1. …that because of the time you spend with your child with DBMD, you don't have enough time for yourself?	0	1	2	3	4
2. …stressed between caring for your child with DBMD and trying to meet other responsibilities (work/family)?	0	1	2	3	4
3. …angry when you are around your child with DBMD?	0	1	2	3	4
4. …that your child with DBMD currently affects your relationship with family members or friends in a negative way?	0	1	2	3	4
5. …strained when you are around your child with DBMD?	0	1	2	3	4
6. …that your health has suffered because of your involvement with your child with DBMD?	0	1	2	3	4
7. …that you don't have as much privacy as you would like because of your child with DBMD?	0	1	2	3	4
8. …that your social life has suffered because you are caring for your child with DBMD?	0	1	2	3	4
9. …that you have lost control of your life since your child's diagnosis of DBMD?	0	1	2	3	4
10…uncertain about what to do about your child with DBMD?	0	1	2	3	4
11…you should be doing more for your child with DBMD?	0	1	2	3	4

12…you could do a better job in caring for your child with DBMD?	0	1	2	3	4

SECTION J: Control

Instructions: This section asks you about how much control you or others have over certain aspects of your child's DBMD. Marking 0 on the scale means you feel that you have no control over a particular aspect, while marking a 10 means you feel that you have complete control, and marking a 5 means you have a medium amount of control.
Please answer each item.

1. In general, how much control do you feel you have over your child's DBMD?

No Control	0	1	2	3	4	5	6	7	8	9	10	Complete Control

2. How much control do you feel you have over your child's daily symptoms?

No Control	0	1	2	3	4	5	6	7	8	9	10	Complete Control

3. How much control do you think you have over the long-term course of your child's DBMD?

No Control	0	1	2	3	4	5	6	7	8	9	10	Complete Control

4. How much control do you think you have over the medical care and treatment of your child's DBMD?

No Control	0	1	2	3	4	5	6	7	8	9	10	Complete Control

5. How much control do you think that others (a spouse, doctor, God, etc.) have over your child's DBMD?

No Control	0	1	2	3	4	5	6	7	8	9	10	Complete Control

SECTION K: Adaptation

Instructions: This section is about the impact that having a child with DBMD has on you. Read and answer all the statements, even if you are not completely sure about your answer. For each statement choose the number that best described how much you agree.

This is what the numbers mean:
1. *Not at all*
2. *A little bit*
3. *Somewhat*
4. *Quite a bit*
5. *Very much*

Being a parent to an individual with Duchenne or Becker muscular dystrophy has...

	Not At All	A Little Bit	Somewhat	Quite a Bit	Very Much
Helped me accept the way things work out	1	2	3	4	5
Helped me learn to deal better with uncertainty	1	2	3	4	5
Taught me how to adjust to things I cannot change	1	2	3	4	5
Helped me take things as they come	1	2	3	4	5
Helped me to look at things in a more positive way	1	2	3	4	5
Helped me learn to handle difficult times	1	2	3	4	5
Helped me become more comfortable with who I am	1	2	3	4	5
Helped me become a stronger person	1	2	3	4	5
Helped me feel better about my ability to handle problems	1	2	3	4	5
Helped me become a better person	1	2	3	4	5

	Not At All	A Little Bit	Somewhat	Quite a Bit	Very Much
Helped me know who I can count on in times of trouble	1	2	3	4	5
Makes me more willing to help others	1	2	3	4	5
Helped relationships become more meaningful	1	2	3	4	5
Helped me become closer to people I care about	1	2	3	4	5
Helped me become more aware of the love and support available from other people	1	2	3	4	5
Helped me learn my life is more meaningful	1	2	3	4	5
Given me a greater appreciation for life	1	2	3	4	5
Helped me develop a deeper sense of purpose in life	1	2	3	4	5
Helped me feel peaceful	1	2	3	4	5
Helped me find strength in my faith or spiritual beliefs	1	2	3	4	5

SECTION L: Update on Your Demographics

1. What is your current marital status?
   o Single, never married
   o Married or long-term committed relationship
   o Separated
   o Divorced
   o Widowed

2. What is your current employment status?
   a. All of my time is spent caring for my family and my home
   b. I have a **full-time** job or schooling program
   c. I have a **part-time** job or schooling program

3. What is your household income?
   o Less than $50,000
   o $50,000 - $99,999
   o $100,000 - $149,999
   o $150,000 - $199,999
   o $200,000 - $249,999
   o $250,000 or more

FOR NEWLY RECRUITED PARTICIPANTS ONLY:

4. What is your year of birth? _____

5. What is your race/ethnicity?
   a. Caucasian/White
   b. African American
   c. Asian/Pacific Islander
   d. Native American
   e. Other

6. Do you consider yourself to be of Hispanic origin
   a. Yes
   b. No

7. What is your highest level of education that you have completed?
   a. Grade School or Middle School
   b. High school diploma/GED or high school equivalent
   c. Some college or technical training
   d. College degree
   e. Post-baccalaureate degree

8. What state do you live in? _____

9. Are you a carrier for DMBD?
   a. Yes, I am a carrier
   b. No, I am not a carrier or I am extremely unlikely to be a carrier
   c. I do not know if I am a carrier

Thank you for completing the survey. If you have any questions or concerns, email Holly Peay at holly@parentprojectmd.org or 443-791-5927, or Megan Bell at bellme@mail.nih.gov. Please expect to receive the next survey in about 12 months. You will notice that some of the questions in the next survey are the same, which allows us to look for change over time.

If you move or change your email address or phone number, please let Holly know. We appreciate your continued involvement in this project.

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National Human Genome Research Institute, NIH, Bethesda, MD
- Sc. M. in Genetic Counseling, January 2015
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Bethel University, St. Paul, MN
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Pediatric Cancer Genetic Counseling Intern, November 2014-December 2014
Department of Genetics, Children's National Medical Center, Washington, D.C.
- Discussed personal and family implications of undergoing genetic testing
- Guided families through the decision-making process
- Generated tailored follow-up patient note which included personalized risks and health information for patients and their families

Internal Medicine Genetic Counseling Intern, September 2014-October 2014
Johns Hopkins Hospital, Baltimore, MD
- Obtained medical intakes, established goals, and contracted with patients
- Counseled patients about common multifactorial disease, including high blood pressure, diabetes, obesity, high cholesterol, and heart disease.

Clinical Research Genetic Counseling Intern, June-July 2014
National Eye Institute; National Institute of Allergy and Infectious Diseases, NIH, Bethesda, MD
- Communicated goals and purposes of research protocols to potential participants
- Provided psychosocial support and identified local and national support organizations for patients and families

Pediatrics Genetic Counseling Intern, March 2014-May 2014
Department of Neurology, Kennedy Krieger Institute, Baltimore, MD
- Consented patients and families for clinical whole exome sequencing
- Researched, annotated, and returned primary and secondary clinical whole exome sequencing variants to families

Clinical Research Genetic Counseling Intern, September 2013-March 2014
National Human Genome Research Institute, National Institutes of Health, Bethesda, MD
- Obtained informed consent for participants in whole genome sequencing research protocols
- Provided comprehensive information about whole genome primary and secondary results to patients and families

Cancer and Prenatal Genetic Counseling Intern, June 2013-August 2013
Centracare Genetics, Centracare Health, St. Cloud, MN
- Performed risk assessment and counseling for familial cancer patients
- Presented and discussed appropriate genetic testing options

- Identified and provided referrals for additional medical and psychosocial care

Prenatal Genetic Counseling Intern, October 2013-May 2013

Center for Maternal and Fetal Medicine, Howard County Medical Center, Columbia, MD

Prenatal Genetic Counseling and Diagnostic Center, Johns Hopkins, Baltimore, MD

- Obtained medical history and constructed pedigrees
- Provided risk assessment and presented genetic testing options to women and families at low and high risk for genetic diseases

INTERNSHIP EXPERIENCE

NYMAC Collaborative Region Genetics and Newborn Screening Fellow, *April 2013-May 2014*

- Gain understanding and perspective on federal policies and programs related to newborn screening and other genetic services
- Participated in SACHDNC Meetings, April 2013 and September 2013

Programs Intern, *September 2011-January 2012*

Genetic Alliance, Washington, DC

- Generated content for newborn screening condition pages on the *Baby's First Test* website
- Collaborated with partner agencies to compile a companion document to the *Healthy People 2020* objectives focused on children and youth with special healthcare needs

RESEARCH EXPERIENCE

Master of Science Thesis, *Where does hope fit in? The relationship between hope, uncertainty, and coping efficacy in mothers of children with Duchenne/Becker Muscular Dystrophy*

- Designed and conducted a unique survey study
- Submitted to and received approval from the NHGRI IRB
- Recruited and consented participants

Laboratory Research Assistant, *June-August 2010, June-August 2009*

Cancer Biology, Sanford Research, Sioux Falls, SD

- Investigated the molecular characteristics of cell signaling pathways in head and neck cancer

PRESENTATIONS

Final Thesis Presentation, NIH, *Where does hope fit in? The relationships among hope, uncertainty, and coping efficacy in mothers of children with Duchenne/Becker Muscular Dystrophy?*, January 16, 2015

Presentation, Post Clinic Conference, NHGRI, *The Role of Family History Information in Motivating At-Risk Individuals To Lifestyle Behavior Change*, October 29, 2014

Abstract and Oral Presentation, NSGC Conference 2014, *Evaluation of Quality and Literacy of Commercial Non-Invasive Prenatal Test (NIPT) Websites*, September 2014

Poster, NHGRI Retreat, *Where does hope fit in? The relationships among hope,*

uncertainty, and coping efficacy in mothers of children with Duchenne/Becker Muscular Dystrophy (A proposed study), December 13, 2013

Abstract, NSGC Conference 2013, *Intimate Partner Violence During Pregnancy: The Role of the Prenatal Genetic Counselor*, October 2013

Presentation, Post Clinic Conference, NHGRI, *Domestic Violence and Pregnancy: Screening, Barriers, and Interventions*, April 17, 2013

PUBLICATIONS

Megan Bell, Sharon F. Terry, Vaughn Edelson. For Fun and Fitness. Exceptional Parent. June 2012. P 31-32

Megan Bell, Sharon F. Terry, Vaughn Edelson. Disability and the Arts: A Palate of Possibilities. Exceptional Parent. March 2012. P. 52-54.

Vermeer PD, **Bell M,** Lee K, Vermeer DW, Ganesan S, Bilal E, Bhanot G, Klingelhutz AJ, Hendriks WJ, Lee J. (2012) ErbB2, EphrinB1, Src Kinase and PTPN13 Signaling Complex Regulates MAP Kinase Signaling in Human Cancers. PLoS Biology.

Storlie EW, Ihry RJ, Baehr LM, Tieszen KA, Engbers JH, Anderson-Daniels JM, Davis EM, Gilbertson AG, Harden NR, Harris KA, Johnson AJ, Kerkvleit AM, Moldan MM, **Bell ME**, and Wanous MK. (2009) Genomic regions influencing gene expression of the HMW glutenins in wheat. Theoretical and Applied Genetics 118(2): 295-303.