THE PSYCHOLOGICAL EFFECTS OF EHLERS-DANLOS SYNDROME: INSIGHTS INTO LIFE WITH AN INVISIBLE ILLNESS

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ABSTRACT

Ehlers-Danlos syndrome (EDS) is a heritable connective tissue disorder caused by an irregularity in the collagen and has complications related to joint hypermobility, skin flexibility, and tissue fragility, (March, 2016). The syndrome is also considered an invisible illness. Because it is not readily apparent. EDS symptoms can affect multiple areas of the body which can lead to a negative prognosis. The diagnosis of EDS, the symptoms, and the prognosis can lead the patient to feelings of psychological distress. The purpose of this study is to explore the prevalence of psychological symptoms in individuals with Ehlers-Danlos syndrome and to also assess the treatment they receive from their peers, family members, school, work, and in medical situations. The study was conducted using a survey on websites established for persons with EDS and included questions assessing anxiety, with the Beck Anxiety Inventory (Beck, Fergus, Valentiner, Gillen, Hiraoka, Twohig, & McGrath, 2012), questions assessing depression, with the Patient Health Questionnaire (Kroenke, Spitzer, & Williams, 2001), and questions assessing treatment from others related to this invisible illness. Although the data gathered are not representative of the EDS population, comparisons with established norms for anxiety and depression are made. In addition, concerns related to treatment from others related to having an invisible illness are discussed.

I. INTRODUCTION

Suffering from a chronic illness is an unfortunate burden that many people around the world experience. More than 117 million adults suffer from one or more chronic illnesses (Ward Schiller, & Goodman, 2014), and millions of those chronic illnesses are not always immediately detected by the doctors. Some of these individuals suffer from an invisible illness. An invisible illness is not readily apparent. "Invisible illnesses can greatly impact both the physical and mental health of individuals. These illnesses are debilitating, preventing participation in the normal activities of daily living. Examples include…Ehlers–Danlos syndrome" (Pederson & Hochstetler, 2016, p. 57). Ehlers-Danlos syndrome (EDS) is a heritable connective tissue disorder caused by an irregularity in the collagen and has complications related to joint hypermobility, skin flexibility, and tissue fragility (March, 2016). EDS can affect many different parts of the body such as the skin, joints and multiple organs. The impact of EDS can be mild to life threatening, depending on the severity of the symptoms.

With the many different parts of the body being affected by Ehlers-Danlos syndrome, the symptoms can be extensive. Symptoms affecting joints can include: joint pain, joint hypermobility, joint swelling, and dislocations (Alan, 2016). This can be very painful for EDS patients. Symptoms affecting the skin can include: easily bruising, slow and poor healing of skin wounds and skin tearing. Since EDS affects the body's biggest organ (the skin), it also affects other organs as well, such as: the eyes, lungs, blood vessels, heart, gastrointestinal system, and many other organs depending on the severity of EDS (Alan, 2016).

In addition to these symptoms, psychological symptoms have been found with EDS, including depression and anxiety disorders (Hershenfeld et al., 2016). Dr. Samantha Hershenfeld conducted a study to identify a connection between psychiatric disorders and EDS. A psychiatric disorder affects a person's mood or behavior. In Hershenfeld's study 45% of the patients suffered from a psychiatric disorder (Hershenfeld et al., 2016). In the study of why EDS patients were more prevalent in having a psychiatric disorder, Hershenfeld wrote, "The presence of any pain symptom was significantly associated with having a psychiatric disorder" (Hershenfeld et al., 2016, p. 341). Since pain is a symptom that is frequently mentioned, it is evident that pain could play a part in the psychological symptoms.

TYPES OF EHLERS-DANLOS SYNDROME

There are three major categories that compromise Ehlers-Danlos syndrome. These are, hypermobility, classical, and vascular type. Each type has its own unique set of symptoms and complications.

Dr. Yael Gazit defines the hypermobility type as the "most common subtype of EDS and the least severe one" (Gazit, Jacob, & Grahame, 2016, p. 1). The most common symptoms are muscle and skeletal pain, joint instability, subluxations (a partial dislocation), and dislocations. One of the main issues with this type, is the difficulty of diagnosis partly because there is not an associated genetic mutation that is connected to hypermobility type. Since there is not an associated genetic mutation, the diagnosis is based on symptoms and the Beighton score. The Beighton score is a nine point system that quantifies the joint laxity and hypermobility (Beighton & Horan, 1969, p. 445). Many doctors do not automatically perceive the symptoms as being EDS and therefore

leave the patient undiagnosed for longer than needs be. This leaves hypermobility type "under recognized and inadequately managed, leading to neglect of these patients, which may lead to severe disability that almost certainly could have been avoided" (Gazit, Jacob, & Grahame, 2016, p. 1-2). Since the patient is left undiagnosed and untreated, feelings of hopelessness and frustration can occur. This can include the patient's feelings of "depression and a lower quality of life that limit their lifestyles, as well as affecting their psychological well-being" (Ainsworth & Aulicino, 1993, p. 252).

The classical type EDS is best characterized by the mutation in the COL1A1 or COL5A1 gene (Nuytinck et al., 2000). Dr. Nuytinck found that the most common symptoms are "skin hyper elasticity, increased tendency to bruise, and abnormal scarring" (Nuytinck et al., 2000, p. 1). Classical type can be difficult to diagnose as well. Not all patients who are genetically tested for this type have a mutation on the COL1A1 or COL5A1 gene. Sara Whitelaw (2003) found that since genetic testing is not fully equipped, this can be a factor because "at present, there is no highly sensitive genetic testing available for the Classical type EDS" (Whitelaw, 2003, p. 425). Since genetic testing for classical type is not completely accurate, the Beighton score is used to assist with the diagnosis (Beighton & Horan, 1969, p. 445). Not having a clear diagnosis can increase depressed feelings in patients. The current lack of genetic knowledge, has patients anxiously waiting to learn more about their disease so they can treat their symptoms more effectively.

Vascular type EDS is the most severe case and is considered life threatening. Dr. Masuno found that this type has a mutation on the COL3A1 gene and has severe symptoms that can result in "arterial, intestinal, and uterine rupture" (Masuno et al., 2012,

p. 207). Vascular type is the easiest to diagnose because of the distinct characteristics that the patients possess, such as varicose veins and translucent skin (Masuno et al., 2012). The genetic testing for this type of EDS has been fairly successful in most patients in terms of finding the correlating gene mutation. Because of the increased risk that vascular EDS has on its patients, the levels of anxiety increases as well. Many of these patients fear the future and suffer from a low quality of life. Jeanna Ford reported that many of these patients fear "the unknown, death and dying, or isolation...All of these feelings can escalate and exacerbate distressing emotions" (Ford, 2016, p. 132). The distressing emotions that can occur can increase the patients' chances of suffering from an anxiety disorder or depression.

DAILY TRIALS

Everyday life for Ehlers-Danlos syndrome patients can be complicated with the daily struggles they encounter. Chronic and constant pain can be one of them. Pain can make daily life more difficult because of the limitations on activities that occur in accordance with the pain (Berglund, Nordström, & Lützèn, 2000). Pain does not only limit the activities that occur during the day, but also at night. Sleeping has been problematic in many EDS patients. Pain keeps patients up at night and they are left exhausted the next day, which can result in limiting daily activities (Berglund, Nordström, & Lützèn, 2000). This can become a cycle. A lack of sleep can cause chronic fatigue that is often found in EDS patients. Nicol Voermans wrote that "severely fatigued patients were more impaired than non-severely fatigued patients and reported a higher level of psychological distress" (Voermans & Knoop, 2011, p. 706). The daily trials of EDS patients makes it difficult for them to do simple tasks such as cleaning and going to

work or school. Patients have also discussed that the daily symptoms (pain, dislocations, and fatigue) that occur, prevent them from striving for a goal or a certain career path (Berglund, et al., 2000). Even though there are certain treatments that can help manage some of these symptoms, many patients seem to deal with these tribulations on a daily basis.

TREATMENTS

Ehlers-Danlos syndrome is associated with many different symptoms that can be managed with the use of different treatment tactics. Treatments include physical therapy to strengthen the muscles around the loose joints and ligaments, braces and splints to provide stability to joints, pain medications, and surgery for the treatment of complications brought on by EDS (March, 2016). The goal of using these treatments is to reduce the symptoms that the patients are having and give the patients a better quality of life.

Lies Rombaut discussed the findings from those who participated in these medical treatments. From the population that was surveyed, Rombaut found that 92.4% of patients used medications, 70.9% went through surgery, and 51.9% were enrolled in physical therapy (Rombaut et al., 2011). Patients expect a positive outcome when participating in these treatments; however, that is not always the case. “Only 33.9% of the patients who underwent surgery and 63.4% of patients in physical therapy reported a positive outcome” (Rombaut et al., 2011, p. 1106). With symptoms not being successfully treated, the pain and other complications continue. This can result in increased psychological symptoms and a feeling of having a low-quality of life. Brita Berglund’s questionnaire study, found that “individuals with Ehlers-Danlos syndrome

symptoms of probable anxiety and depression were commonly reported" (Berglund, Pettersson, Pigg, & Kristiansson, 2015, p. 4). Unsuccessful treatment for Ehlers-Danlos syndrome patients increase the number of psychological symptoms that occur and can make managing this syndrome challenging.

PSYCHOLOGICAL DISTRESS

The diagnosis, prognosis, symptoms, and social situations related to Ehlers-Danlos syndrome, can result in increased psychological distress in patients with Ehlers-Danlos syndrome. Many patients are suffering with symptoms for an extended period of time before they are diagnosed. Although having a diagnosis can be relieving to people because their symptoms are validated, many people experience distress because of the diagnosis. Baeza-Velasco found that Ehlers-Danlos syndrome "patients often have negative expectations about their own ability" (Baeza-Velasco, Gely-Nargeot, Vilarrasa, & Bravo, p. 1132). This way of thinking increases depressive thoughts about the diagnosis of their condition (Baeza-Velasco, Gely-Nargeot, Vilarrasa, & Bravo). EDS cannot be cured and patients can feel distressed knowing that their symptoms will never fully cease.

The prognosis that follows the diagnosis can be unsettling to patients because it could determine their life expectancy. Since vascular EDS is more life threatening, the prognosis could be devastating to patients and can result in them developing depression and anxiety. Lawrence found that prognosis' can also involve trouble with pregnancy and delivery, increased loss of mobility, and death (Lawrence, 2005). Many different prognoses are negative, and that results in patients thinking of the unknown; this can increase anxiety. "Fear leads to avoidance behaviors that create a vicious circle which

contributes to worsening the scenario; a person who has pain, especially on movement, tends to avoid doing things that provoke their symptoms" (Baeza-Velasco, et al., p. 1132). Having fear can lead to a decrease of daily activities that can be disadvantageous. The idea that the patient's quality of life is going to decrease is troublesome and can result in psychological distress.

Patients also feel a lack of understanding from their peers. EDS patients find that many people do not believe someone is disabled if they do not visibly see it. This can be difficult if the patient is in need of certain accommodations. Davis found that "those whose disabilities are invisible may also have to convince other people that they really are disabled, not seeking…advantage: thus, what they must do is meet a burden of proof. They thus face a double bind: either they forgo the assistance or accommodation they need…or they endure the discomfort of subjecting themselves to strangers' interrogations" (Davis, 2005, p. 154). The lack of acknowledgment of this invisible disease can cause psychological distress to the patients because they have to provide proof that they need assistance. As a consequence of this, patients will forgo the assistance that they need, or endure the insensitivity towards their disability. This increases the association of psychological distress and can increase the probability of a psychiatric disorder (Murray, et al., 2013).

Ehlers-Danlos syndrome patients find it difficult to find understanding from not only personal relationships, but from medical professionals as well. EDS is a rare genetic disorder than many people have not heard of, which could result in a lack of compassion and sincerity when in contact with EDS patients. Patients feel that their symptoms are not taken seriously when they are being treated. Murray found that the diagnosis of EDS, is

"often associated with depression and anxiety that is amplified by a lack of recognition and knowledge of the syndrome by managing clinicians" (Murray, Yashar, Uhlmann, Clauw, & Petty, 2013, p. 2982). This can increase the chances of the patient not being treated medically as they should.

COPING

Different coping mechanisms can be beneficial to Ehlers-Danlos syndrome patients if they are experiencing psychological distress. Since EDS is a chronic illness, many people seek out other treatments that will decrease the feelings of depression and anxiety. One option that can be beneficial is cognitive therapy. Attending therapy with a trained counselor, can provide the patient with coping mechanisms in order to deal with the psychological effects that result from their illness. The therapist can help "explore the motivation and the expectations of patients regarding a psychological intervention" (Baeza-Velasco, et al., p. 1135). The therapist can help the patient receive the treatment that they seek. Therapy "showed demonstrable improvements in…mood and self-efficacy were found with notable reductions in pain-related 'catastrophizing' and disability and fear" (Baeza-Velasco, et al., p. 1133). These coping mechanisms can increase the quality of life in patients that suffer from the psychological effects of EDS.

Another coping mechanism that patients have found to ease their psychological distress is meditation. Williams-Orlando found that "meditation is mental exercise for the purpose of clearing, quieting, centering, or focusing the mind; reducing stress; or reaching heightened levels of spiritual awareness" (Williams-Orlando, 2012, p. 36). Many patients believe that this can decrease the psychological effects associated with their illness. Patients have noticed that "meditation has demonstrated (1) positive effects

on both the brain and immune system functioning, (2) increased positive coping and ability to relax, (3) reduced pain and anxiety, (4) prevention of relapse in depression and substance abuse, and (5) improvement in chronic insomnia" (Williams-Orlando, 2012, p. 37). Some patients have found alternative ways to treat their psychological distress besides medications, and those patients have found these methods to have a positive effect on their lives.

PURPOSE OF THE STUDY

The research done on the psychological effects of Ehlers-Danlos syndrome, has shown an increased psychological effect on EDS patients related to diagnosis, prognosis, symptoms (pain, fatigue, dislocations, etc.), and social situations. There has been little research done on the treatment that EDS patients face from their peers and physicians. Thus, I will be expanding the knowledge of the treatment that EDS patients face by adding to the proceeding research.

I will be conducting a research study with IRB approval in the hopes of finding what psychological effects occur in Ehlers-Danlos syndrome. I will be focusing on the psychological disorders, anxiety and depression to see if there are any links between those psychological disorders and EDS. The sample will be taken from online Ehlers-Danlos Syndrome support groups and will be statistically analyzed to see if the results are significant or not. The research study will be conducted in a survey form that will be easily accessible to the sample. In order to measure the anxiety and depression that is found in EDS patients, I will be using the Beck Anxiety Inventory (BAI) (Beck et al., 2012) and the Patient Health Questionnaire (PHQ-9) (Kroenke K, Spitzer R L, Williams J B, 2001)

RESEARCH QUESTIONS

The BAI scoring will be based on the calculation of the twenty one questions that are listed. A score of 0 – 21 = low anxiety, a score of 22 – 35 = moderate anxiety, and a score of 36 and above = potentially concerning levels of anxiety (Beck et al., 2012). The values of the answers are not equivalent. The answer "not at all" is 0 points, "mildly - but it didn't bother me much" is 1 point, "moderately - it wasn't pleasant at times" is 2 points, and "severely – it bothered me a lot" is 3 points (Beck et al., 2012). With the use of the BAI, I will be able to appropriately scale the anxiety that the EDS patient is experiencing.

The PHQ-9 scoring will be based on the calculation of the nine questions that are listed and one follow up question. A score of 0 – 4 = minimal depression, a score of 5 – 9 = mild depression, a score of 10 – 14 = moderate depression, a score of 15 – 19 = moderately severe depression, and a score of 20 – 27 = severe depression. The scoring of the questions will be based on the number of times a participant chooses the answers: "not at all", "several days", "more than half the days", or "nearly every day". Then, the amount of times a participant chooses an answer, it will be multiplied by the number associated with the answer choice. Not at all would be multiplied by 0, several days would be multiplied by 1, more than half the days would be multiplied by 2, and nearly every day would be multiplied by 3. Lastly, the scores will be added together to reveal the final score (Kroenke K, Spitzer R L, Williams J B, 2001). With the use of the PHQ-9, I will be able to appropriately scale the depression that the EDS patient is experiencing.

I will also be conducting research on the perception EDS patients have with their treatment by peers and clinicians in regards to their invisible disease, Ehlers-Danlos Syndrome. In this third part of the survey, the questions will be geared towards the EDS

patients' feelings towards their treatment from peers, family members, school, work, and in medical scenarios. I will be using a scale with the answers: "never", "rarely", "neutral", "often", "always", in order for the participants to accurately describe the feelings they have in regards to the questions being asked. This portion of the survey has little research background and this will add to the better understanding of the psychological effects of Ehlers-Danlos Syndrome.

II. METHOD

PARTICIPANTS AND PROCEDURE

A sample of 385 Ehlers-Danlos syndrome support group members participated in this study by completing an online survey through Qualtrics. The participants were predominately female (96.3%) with the majority being in the 30-40 age group (26.5%, range 18-50 and above). 93.2% of participants identified their ethnicity as Caucasian, 0.3% identified as African American, 2.3% identified as Hispanic/Latin American, 0.3% identified as Asian, and 3.9% identified themselves as Other. Socioeconomically, 3.7% of participants identified themselves as Upper Class, 24.3% identified as Upper-Middle Class, 42.8% identified as Middle Class, 20.9% identified as Lower-Middle Class, and 8.4% identified as Lower Class.

MEASURES

The online survey (see Appendix) given to each participant contained the following four measures: (1) Demographic Information, (2) The Beck Anxiety Inventory (BAI) (3) The Patient Health Questionnaire (PHQ-9), and (4) The Ehlers-Danlos Syndrome Questionnaire.

DEMOGRAPHIC INFORMATION

Participants completed a demographic questionnaire, which included questions regarding the participant's sex, age, ethnicity, and socioeconomic status.

THE BECK ANXIETY INVENTORY (BAI)

The Beck Anxiety Inventory (BAI) (Beck et al., 2012) consists of twenty-one questions that measure the severity of the participant's anxiety levels. Each question has four responses: "not at all", "mildly - but it didn't bother me much", "moderately - it wasn't pleasant at times", and "severely – it bothered me a lot". The BAI has a maximum score of sixty-three.

PATIENT HEALTH QUESTIONNAIRE (PHQ-9)

The Patient Health Questionnaire (PHQ-9) (Kroenke K, Spitzer R L, Williams J B, 2001) consists of nine questions and one follow up question that measure the severity of the participant's depression levels. Each question has the following four responses: "not at all", "several days", "more than half the days", or "nearly every day". The PHQ-9 has a maximum score of twenty-seven.

EHLERS-DANLOS SYNDROME QUESTIONNAIRE

The Ehlers-Danlos Syndrome questionnaire consists of one preliminary question and eleven questions set to a scale that measure the treatment the Ehlers-Danlos syndrome patients feel they receive from their peers, family members, at school, at work, and in medical situations. Each question has the following five responses: "never", "rarely", "neutral", "often", "always". This portion of the survey is being used for the first time in this research and will contribute to the better understanding of the psychological effects of Ehlers-Danlos syndrome.

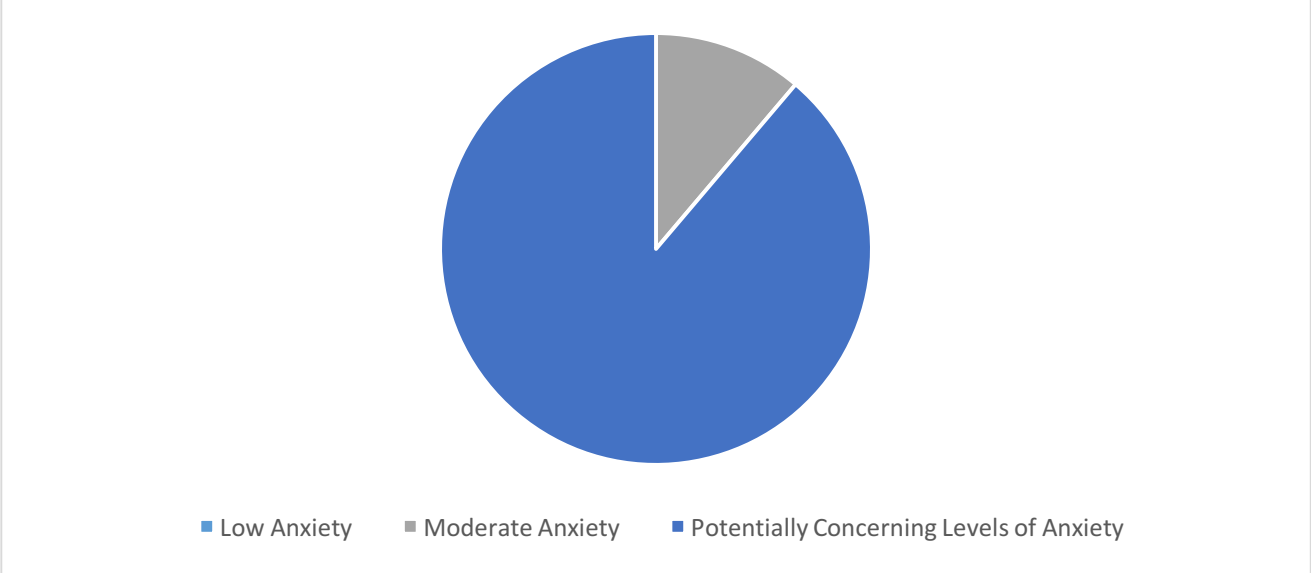
III. RESULTS

BECK ANXIETY INVENTORY (BAI)

The Beck Anxiety Inventory (BAI) section of the survey, exhibited high ranges of anxiety from the participants (see Chart 1). After scoring the participants answers, it was revealed that 0% of participants answered in the mild anxiety range, 11.2% of participants answered in the moderate anxiety range, and 88.8% of participants answered in the potentially concerning levels of anxiety range.

Chart 1

Beck Anxiety Inventory (BAI) Ranges



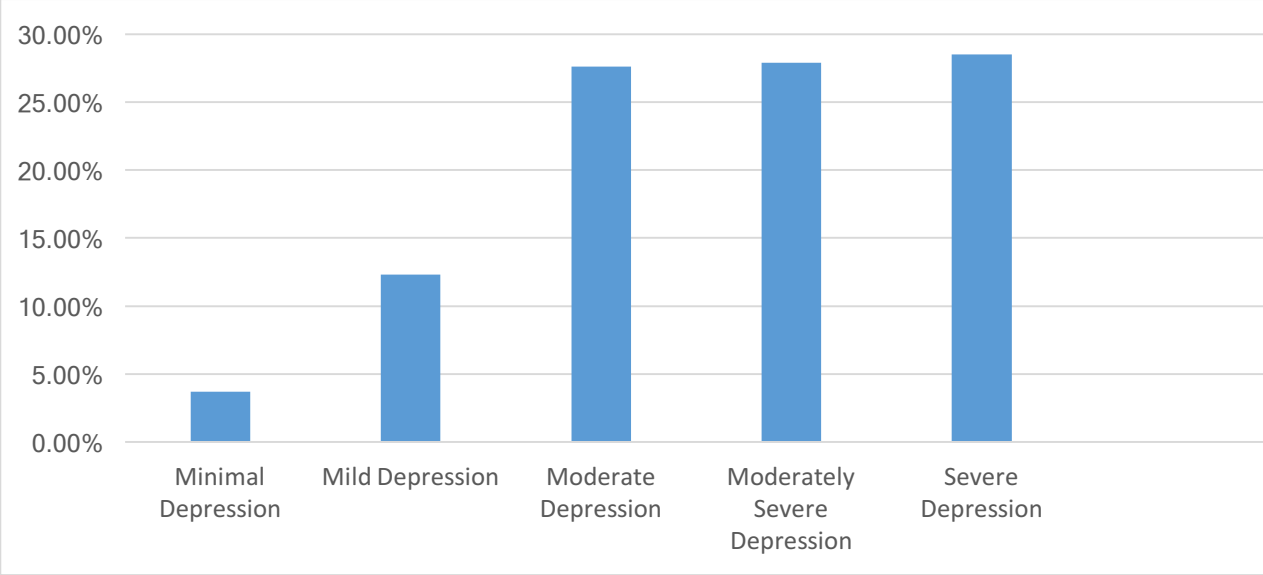
PATIENT HEALTH QUESTIONNAIRE (PHQ-9)

The Patient Health Questionnaire (PHQ-9) section of the survey, exhibited high ranges of depression from the participants (see Chart 2). After scoring participants answers, it was revealed that 3.7% of participants answered in the minimal depression range, 12.3% of participants answered in the mild depression range, 27.6% of participants

answered in the moderate depression range, 27.9% of participants answered in the moderately severe depression range, and 28.5% of participants answered in the severe depression range.

Chart 2

Patient Health Questionnaire (PHQ-9) Ranges



EHLERS-DANLOS SYNDROME QUESTIONNAIRE

The Ehlers-Danlos syndrome questionnaire (see Table 1) had twelve questions that participants answered in the survey. Majority of the answers reflected the hypothesis that Ehlers-Danlos syndrome patients encounter negative social interactions with family members, friends, in school, in work, and in medical situations.

Table 1

Ehlers-Danlos Syndrome Questionnaire

	YES	NO
Do you use a handicap spot?	43.6%	56.4%

	NEVER	RARELY	NEUTRAL	OFTEN	ALWAYS
If the answer is yes on question #36, have you ever received a dirty look from someone when using a handicap spot?	20.8%	22.4%	16.9%	35.0%	4.9%
If the answer is yes on question #36, have you ever received a negative comment from someone when using a handicap spot?	44.6%	28.0%	12.5%	13.7%	1.2%
Do you feel your Ehlers-Danlos Syndrome symptoms are taken seriously in medical situations?	9.1%	52.7%	22.5%	14.2%	1.4%
Do you feel that your medical care is compromised because of the lack of understanding towards your Ehlers-Danlos Syndrome symptoms?	1.1%	6.6%	9.4%	47.7%	35.1%
Has a medical professional ever commented on the lack of severity towards your Ehlers-Danlos Syndrome symptoms?	14.6%	14.3%	23.4%	42.0%	5.7%
Do you feel your Ehlers-Danlos Syndrome symptoms are taken seriously in your personal life?	7.4%	39.7%	22.3%	27.7%	2.9%
Do you feel that your personal relationships suffer because of the lack of understanding towards your Ehlers-Danlos Syndrome symptoms?	4.0%	13.7%	17.4%	49.6%	15.4%

Has a family member or friend commented on the lack of severity towards your Ehlers-Danlos Syndrome symptoms?	11.5%	23.2%	23.5%	35.2%	6.6%
Do you feel your Ehlers-Danlos Syndrome symptoms are taken seriously in your workplace or school?	16.1%	23.8%	40.5%	18.2%	1.5%
Do you feel that your academics or career suffer because of the lack of understanding towards your Ehlers-Danlos Syndrome symptoms?	4.3%	8.1%	26.9%	33.8%	26.9%
Has an employer, colleague, or a professional from your school (teacher, principal, dean, etc.) commented on the lack of severity towards your Ehlers-Danlos Syndrome symptoms?	24.6%	13.0%	39.3%	19.8%	3.3%

PEARSON CORRELATION

Pearson correlations (see Table 2) were performed among the Beck Anxiety Inventory (BAI), Patient Health Questionnaire (PHQ-9), and the Ehlers-Danlos syndrome questionnaire measures. Although a large number of correlations are reported, many are at the 0.01 level of significance. The PHQ-9 and the BAI were significantly positively correlated. A majority of the EDS questionnaire measures and BAI measures significantly positively correlated with the exception of three of the EDS questionnaire measures resulting in significantly negative correlations. A majority of the EDS questionnaire measures and PHQ-9 measures significantly positively correlated with the exception of four of the EDS questionnaire measures resulting in significantly negative correlations.

Table 2

Pearson Correlations Results

	BAI	PHQ-9
BAI	1	.647**
QUESTION 38	.174*	.148
QUESTION 39	-.167**	-.265**
QUESTION 40	.248**	.339**
QUESTION 41	.080	.123*
QUESTION 42	-.116*	-.185**
QUESTION 43	.319**	.420**
QUESTION 44	.243**	.263**
QUESTION 45	-.086	-.182**
QUESTION 46	.410**	.433**
QUESTION 47	.245**	.186**

Note. BAI=Beck Anxiety Inventory, PHQ-9=Patient Health Questionnaire, Question 38-47=correlating Ehlers-Danlos Syndrome Questionnaire (see appendix).

*p<0.05, **p<0.01

IV. DISCUSSION

DISCUSSION OF RESULTS

A central purpose of this study was to add to the limited research available on the psychological effects of Ehlers-Danlos syndrome and the psychological effects from the social treatments EDS individuals receive. There still remains a lack of research on this topic to fully comprehend how these situations truly effect Ehlers-Danlos syndrome patients.

One of the essential research questions was to answer if there was a relationship between Ehlers-Danlos syndrome patients and anxiety. The majority of the participants scored in the potentially concerning levels of anxiety range. Since there was a significant amount of scores in the potentially concerning levels of anxiety range, this showed a significant correlation between Ehlers-Danlos syndrome patients and anxiety. This aligns with the previous research that has been completed regarding the induced anxiety that EDS patients can face (Baeza-Velasco, et al., p. 1132).

The next primary research question was to determine if there was a relationship between Ehlers-Danlos syndrome patients and depression. Comparable to the results from measuring anxiety, the majority of participants scored in the severe depression range. Considering the significant amount of scores in the severe depression range, this proves a significant correlation between Ehlers-Danlos syndrome patients and depression. The previous literature on EDS and depression (Berglund, Pettersson, Pigg, &

Kristiansson, 2015, p. 4), coordinates with the findings that I have found in regards to depression and Ehlers-Danlos syndrome patients.

After determining the anxiety and depression of the Ehlers-Danlos syndrome individuals, my next set of research questions was to determine how EDS patients felt about the treatments they received from their peers, family members, in school, in work, and in medical situations. These set of research questions started with a preliminary question regarding if the participant used a handicap parking spot. Although majority of the participants did not utilize a handicap parking spot, many of the participants were still affected by the social interactions that were used as examples.

After addressing the question regarding if the participant utilized a handicap spot, the questions were then based on a scenario that includes the reactions that occurred after the participant used their handicap spot. Many of the participants received more dirty looks than comments from a bystander. However, these reactions that the Ehlers-Danlos syndrome patients receive induces the probability of psychological distress. These research questions identifies that EDS individuals are in fact receiving these reactions.

The next set of questions were relating to the treatment the participants received in medical situations. There was a significant amount of answers that correlated with negative interactions in medical situations. These negative interactions indicate that there is a lack of understanding that is coming from medical personnel and with these negative interactions, there is a potential increase in anxiety, depression, and other psychological disorders. These findings correlates with previous research that has been completed on negative medical situations and Ehlers-Danlos syndrome individuals (Murray, Yashar, Uhlmann, Clauw, & Petty, 2013, p. 2982)

Afterwards, the research questions pertained to the treatment the participants received from their personal relationships. There was a significant amount of responses that associated with negative interactions accompanying their personal relationships. These negative interactions demonstrates the lack of understanding and knowledge from their peers and family members. As a result of this, there is a possibility of an increase in psychological distress in Ehlers-Danlos syndrome patients. The responses that the participants had regarding their negative interactions with their personal relationships, aligns with the previous research found on the psychological effects that can occur because of the lack of understanding towards their invisible illness from their peers and family members (Davis, 2005, p. 154).

Then, the research questions referred to the treatment the participants received in school and in work situations. The responses from the participants showed that there was a significant amount of negative interactions in school and in work situations. In previous literature, there is a lack of knowledge of Ehlers-Danlos syndrome among many individuals because of the rarity of the syndrome. Considering this, many EDS patients suffer from the effects in their work place or in school situations (Davis, 2005, p. 154). The findings from these research questions proves that there is negative interactions in work and in school situations.

Next, I wanted to determine if there was a correlation between anxiety and the Ehlers-Danlos syndrome questions I asked regarding the treatment EDS patients received from their peers, family members, in school, in work, and in medical situations. After completing a Pearson correlation, I have concluded that the majority of participant's answers showed that anxiety and the Ehlers-Danlos syndrome questions were

significantly positively correlated at the 0.01 level of significance. These findings confirmed the increase in anxiety that Ehlers-Danlos syndrome patients can experience from the treatment that they receive from their peers, family members, in school, in work, and in medical situations (Murray, et al., 2013).

Lastly, I wanted to determine if there was a correlation between depression and the Ehlers-Danlos syndrome questions I asked regarding the treatment EDS patients received from their peers, family members, in school, in work, and in medical situations. After completing a Pearson correlation, I have concluded that the majority of participant's answers showed that depression and the Ehlers-Danlos syndrome questions were significantly positively correlated at the 0.01 level of significance. The amount of depression that was found, validated the increase in depression Ehlers-Danlos syndrome patients can encounter from the treatment that they receive from their peers, family members, in school, in work, and in medical situations (Murray, Yashar, Uhlmann, Clauw, & Petty, 2013, p. 2982).

STRENGTHS AND LIMITATIONS

A primary limitation of this research study is that it is not representative of a larger population of Ehlers-Danlos syndrome patients. The sample taken for this research study was only EDS individuals involved in these particular online support groups. The support group members can vary in severity of their symptoms which can also determine if less severe or more severe Ehlers-Danlos syndrome individuals participated in the research study. On account of this, the research study does not reflect Ehlers-Danlos syndrome patients as a whole

Since the online survey was anonymous, it was not possible to verify the participant's diagnosis. As a result of this, the participants could vary in their diagnosis stage and therefore makes it difficult to identify whether or not each participant had an official diagnosis or not.

Majority of the Ehlers-Danlos syndrome support groups that participated included members from multiple areas of the world. This allowed the survey to be world-wide and reach a diverse set of Ehlers-Danlos syndrome individuals.

IMPLICATION FOR FUTURE RESEARCH

The findings of this research study confirmed that there is an occurrence of psychological distress among Ehlers-Danlos syndrome patients because of the treatment they receive in social situations. Although there was significant correlations found between anxiety, depression, and the treatment EDS patients receive, there is more to be found on the psychological effects caused by other scenarios and situations. There is also further knowledge to be found on the other psychological effects that could occur besides anxiety and depression. As a result of this, the continuation of this subject of research needs to occur to advance the education on the psychological effects of Ehlers-Danlos syndrome.

CONCLUSION

Ehlers-Danlos syndrome, an invisible illness, is a rare genetic disorder that many individuals (including medical personnel) do not have the knowledge of. This can result in a lack of understanding from family members, peers, in school, in work, and in medical situations (Murray, et al., 2013). This can also result in poor medical care from

health care providers (Gazit, Jacob, & Grahame, 2016, p. 1-2). With this lack of understanding and the difficulty of coping with this rare genetic disorder, there is an increase in psychological distress among Ehlers-Danlos syndrome patients (Murray, et al., 2013). In the research study that I conducted, I confirmed this previous research with the results of significant correlations between anxiety, depression, and the treatment EDS individuals receive. Although there has been an increase in knowledge from this research study, there is still a significant amount of research that needs to be conducted on the psychological effects of Ehlers-Danlos syndrome.

V. APPENDIX

DEMOGRAPHIC INFORMATION

1. Your sex

    a. Male

    b. Female

    c. Other

2. Age

    a. 18-21

    b. 21-30

    c. 30-40

    d. 40-50

    e. Above 50

3. Ethnicity

    a. Caucasian/White

    b. African American

    c. Hispanic/Latin American

    d. Asian

    e. Other

4. How would you describe your socioeconomic status?

    a. Upper class

    b. Upper-middle class

c. Middle class

d. Lower-middle class

e. Lower class

THE BECK ANXIETY INVENTORY (BAI)

Use the scale below to describe how you feel on items 5-25:

A	B	C	D
Not at all	Mildly-but it didn't bother me much	Moderately-it wasn't pleasant at times	Severely-it bothered me a lot

5. Numbness or tingling

6. Feeling hot

7. Wobbliness in legs

8. Unable to relax

9. Fear of worst happening

10. Dizzy or lightheaded

11. Heart pounding/racing

12. Unsteady

13. Terrified or afraid

14. Nervous

15. Feeling of choking

16. Hands trembling

17. Shaky/unsteady

18. Fear of losing control

19. Difficulty in breathing

20. Fear of dying

21. Scared

22. Indigestion

23. Faint/lightheaded

24. Face flushed

25. Hot/cold sweats

PATIENT HEALTH QUESTIONNAIRE (PHQ-9)

Over the last 2 weeks, how often have you been bothered by any of the following problems?

Use the scale below to describe how you feel on items 26-34

A	B	C	D
Not at all	Several days	More than half the days	Nearly every day

26. Little interest or pleasure in doing things

27. Feeling down, depressed, or hopeless

28. Trouble falling or staying asleep, or sleeping too much

29. Feeling tired or having little energy

30. Poor appetite or overeating

31. Feeling bad about yourself or that you are a failure or have let yourself or your family down

32. Trouble concentrating on things, such as reading the newspaper or watching television

33. Moving or speaking so slowly that other people could have noticed. Or the opposite being so fidgety or restless that you have been moving around a lot more than usual

34. Thoughts that you would be better off dead, or of hurting yourself

Use the scale below to describe how you feel on items 35:

A	B	C	D
Not difficult at all	Somewhat difficult	Very difficult	Extremely difficult

35. If you checked off any problems, how difficult have these problems made it for you to do your work, take care of things at home, or get along with other people?

EHLERS-DANLOS SYNDROME QUESTIONNAIRE

36. Do you use a handicap parking spot?

a. Yes

b. No

Use the scale below to describe how you feel on items 37-43:

A	B	C	D	E
Never	Rarely	Neutral	Often	Always

37. If the answer is yes on question #36, have you ever received a dirty look from someone when using a handicap spot?

38. If the answer is yes on question #36, have you ever received a negative comment from someone when using a handicap spot?

39. Do you feel your Ehlers-Danlos Syndrome symptoms are taken seriously in medical situations?

40. Do you feel that your medical care is compromised because of the lack of understanding towards your Ehlers-Danlos Syndrome symptoms?

41. Has a medical professional ever commented on the lack of severity towards your Ehlers-Danlos Syndrome symptoms?

42. Do you feel your Ehlers-Danlos Syndrome symptoms are taken seriously in your personal life?

43. Do you feel that your personal relationships suffer because of the lack of understanding towards your Ehlers-Danlos Syndrome symptoms?

44. Has a family member or friend commented on the lack of severity towards your Ehlers-Danlos Syndrome symptoms?

45. Do you feel your Ehlers-Danlos Syndrome symptoms are taken seriously in your workplace or school?

46. Do you feel that your academics or career suffer because of the lack of understanding towards your Ehlers-Danlos Syndrome symptoms?

47. Has an employer, colleague, or a professional from your school (teacher, principal, dean, etc.) commented on the lack of severity towards your Ehlers-Danlos Syndrome symptoms?

VI. REFERENCES

Ainsworth SR, Aulicino PL. 1993. *A survey of patients with Ehlers-Danlos syndrome.* Clinical Orthopaedics and Related Research 286:250–256.

Alan, R. (2016). Ehlers-Danlos Syndrome. *Health Library: Evidence-Based Information*.

Baeza-Velasco, C., Gely-Nargeot, M., Vilarrasa, A., & Bravo, J. (n.d). Joint Hypermobility syndrome: problems that require psychological intervention. *Rheumatology International, 31(9),* 1131-1136.

Beck, A. T., Fergus, T. A., Valentiner, D. P., Gillen, M. J., Hiraoka, R., Twohig, M. P., & ... McGrath, P. B. (2012). Beck Anxiety Inventory. *Psychological Assessment*, *24*(2), 402-408.

Beighton, P., Horan, F. (1969). Orthopedic aspects of the Ehlers-Danlos syndrome. *J Bone Joint Surg [Br], 51, 444-453.*

Berglund, B., Nordström, G., & Lützén, K. (2000). Living a restricted life with Ehlers-Danlos syndrome (EDS). *International Journal Of Nursing Studies*, *37*(2), 111-118.

Berglund, B., Pettersson, C., Pigg, M., & Kristiansson, P. (2015). Self-reported quality of life, anxiety and depression in individuals with Ehlers-Danlos syndrome (EDS): a questionnaire study. *BMC Musculoskeletal Disorders*, *16*89. doi:10.1186/s12891-015-0549-7

Davis, N. A. (2005). *Invisible Disability.* Ethics, 116(1), 153-213.

Ford, J. A. (2016). The Complexity of Assessment and Treatment for Anxiety in Patients With a Terminal Illness. *Journal Of Hospice & Palliative Nursing*, *18*(2), 131. doi:10.1097/NJH.0000000000000223

Gazit, Y., Jacob, G., & Grahame, R. (2016). Ehlers-Danlos Syndrome--Hypermobility Type: A Much Neglected Multisystemic Disorder. *Rambam Maimonides Medical Journal*, *7*(4), 1-10. doi:10.5041/RMMJ.10261

Hershenfeld, S. A., Wasim, S., McNiven, V., Parikh, M., Majewski, P., Faghfoury, H., & So, J. (2016). Psychiatric disorders in Ehlers-Danlos syndrome are frequent, diverse and strongly associated with pain. *Rheumatology International*, *36*(3), 341-348. doi:10.1007/s00296-015-3375-1

Kroenke K, Spitzer R L, Williams J B (2001). The PHQ-9: validity of a brief depression severity measure. *Journal of General Internal Medicine*, 16(9): 606-613.

Lawrence, E. (2005). The clinical presentation of Ehlers-Danlos syndrome. *Advances In*

Neonatal Care (Elsevier Science), 5(6), 301-314.

March, P. P. (2016). Ehlers-Danlos Syndrome. *CINAHL Nursing Guide*.

Masuno, M., Watanabe, A., Naing, B. T., Shimada, T., Fujimoto, W., Ninomiya, S., & ... Kuroki, Y. (2012). Ehlers-Danlos syndrome, vascular type: a novel missense mutation in the COL3A1 gene. *Congenital Anomalies*, *52*(4), 207-210. doi:10.1111/j.1741 4520.2011.00353.x

Murray, B., Yashar, B., Uhlmann, W., Clauw, D., & Petty, E. (2013). Ehlers-Danlos syndrome, hypermobility type: A characterization of the patients' lived experience. *American Journal Of Medical Genetics, Part A*, *161*(12), 2981-2988. doi:10.1002/ajmg.a.36293

Nuytinck, L., Freund, M., Lagae, L., Pierard, G., Hermanns-Le, T., & De Paepe, A. (2000). Classical Ehlers-Danlos syndrome caused by a mutation in type I collagen. *American Journal Of Human Genetics, 66(4), 1398-1402.*

Pederson, C. c., & Hochstetler Mayer, G. g. (2016). The tangible effects of invisible illness. *Counseling Today*, 58(11), 56-59.

Rombaut, L., Malfait, F., De Wandele, I., Cools, A., Thijs, Y., De Paepe, A., & Calders, P. (n.d). Medication, Surgery, and Physiotherapy Among Patients With the

Hypermobility Type of Ehlers-Danlos Syndrome. *Archives Of Physical Medicine And Rehabilitation*, *92*(7), 1106-1112.

Voermans, N. C., & Knoop, H. (2011). Both pain and fatigue are important possible determinants of disability in patients with the Ehlers-Danlos syndrome hypermobility type. *Disability And Rehabilitation: An International, Multidisciplinary Journal*, *33*(8), 706-707. doi:10.3109/09638288.2010.531373

Ward, B. W., Schiller, J. S., & Goodman, R. A. (2014, April 17). Multiple Chronic Conditions Among US Adults: A 2012 Update. Retrieved October 31, 2016, from http://www.cdc.gov/pcd/issues/2014/13_0389.htm

Whitelaw, S. (2003). Ehlers-Danlos syndrome, classical type: case management. Pediatric Nursing, 29(6), 423-432.

Williams-Orlando, C. (2012). Spirituality in integrative medicine. *Integrative Medicine: A Clinician's Journal, 11(4), 34-40.*