Living with Ehlers Danlos Syndrome: An Emerging Adulthood Perspective

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ABSTRACT

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A thesis presented to the Graduate Program in Genetic Counseling

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Ehlers Danlos syndrome (EDS) is a multi-systemic, genetic, connective tissue disorder. This study is an early look at the specific impacts of hypermobile and classical EDS in the transitional period of life between the ages of 20-30, coined, emerging adulthood. Participants were recruited through support groups across the United States and internationally, and were asked to complete an online survey designed to collect both quantitative and qualitative data that focused on areas of life that are important to emerging adulthood including education, career planning and relationships. A total of 368 participants expressed varying degrees of adjustment to a diagnosis of EDS. Specific challenges that were highlighted by participants included an impact on relationships with family, friends and romantic partners based on an individual's severity of symptoms, number of symptoms and current age, as well as experiencing negative physical and emotional impacts of EDS. These results suggest the need for medical professionals to support their patients in a multitude of ways and to be sensitive to this time of transition for those with EDS.

Keywords: Ehlers Danlos syndrome, Emerging Adulthood, Transition, Genetic Counseling, Relationships, Emotional Impact, Physical Impact

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INTRODUCTION

Ehlers Danlos Syndrome

Ehlers Danlos syndrome (EDS) is a multi-systemic, genetic, connective tissue disorder. Many manifestations have been described, including cardiovascular, autonomic nervous system, gastrointestinal, hematologic, ocular, gynecologic, neurologic, and psychiatric, emphazing the multi-systemic nature of EDS (Gazit et al., 2016). The thirteen subtypes of EDS are typically characterized by joint hypermobility, dislocations, and chronic pain, ranging in severity and the type of symptoms that arise, with the core of the condition arising from collagen defects. The prevalence is 1 in 2,500 to 1 in 5,000 live births, affecting both males and females across ethnicities. This study focused on individuals with the Hypermobile and Classical types, which are by far the most common (Castori, 2015).

Ehlers Danlos syndrome hypermobility type (hEDS) is the most common EDS subtype, and while it is often considered the least severe form of EDS, it results in significant pain and disability if not managed properly. Individuals with hEDS experience dislocations and joint inflammations, musculoskeletal complaints, soft tissue overuse, and a variety of other manifestations (Gazit et al., 2016). The genetic etiology of hEDS is unidentified.

Ehlers Danlos syndrome classical type (cEDS) is characterized by joint instability, fragile and/or soft skin, and joint hyper-extensibility. About 50% of patients with the classical form of EDS harbor a mutation in COL5A1 or COL5A2, although this may be an underestimate (Malfait

et al., 2010). The long term physical symptoms of hEDS and cEDS may cause emotional and social difficulties that negatively impact the transition to adulthood.

Emerging Adulthood

In the emerging adulthood period of life, individuals have left behind childhood, but have not yet accepted all of the responsibilities of adulthood. The emerging adulthood period of life is a time where young adults, aged 18-29, are exploring different career choices, love interests, and weighing in on their views of the world. Individuals in this time frame have reached both sexual and physical maturity, with many combining work and education (Arnett, 2014). In the United States, it has been observed that individuals often do not consider themselves to be adults until their late twenties (Arnett, 2000; Lopez et al., 2005; Molgat, 2007; Tanner & Arnett, 2011). Some have attributed the expansion of this stage to cultural and economic shifts, such as increasing tendencies to delay marriage, and rising numbers of young adults seeking tertiary education (Arnett, 2000; Cote, 2000, 2006; Douglass, 2007).

Recent data shows that modern adults put off marriage and parenthood until around age 30 (Arnett 2014). Therefore, the third decade of life has shifted from a focus on starting a family to exploring other life directions such as education and careers (Shulman & Connolly, 2013; Arnett 2014). With this shift, individuals are seeking careers that can support a family as well as the individual. In a study conducted by Lopez et al. 2005, the only general theme that arose out of 118 participants was entry into the workforce and career development. These two aspects of maturity correlate with entering adulthood. Entering the workforce allowed for positive feelings by experiencing happiness within a career, making a living, and working hard towards a goal

(Lopez et al., 2005). However, the importance of a career is often offset by continuing education.

According to Arnett (2000), school attendance is another area in which there is substantial change and diversity among emerging adults. After completion of high school, over half of individuals enter college to further their education. Continuing education at the graduate level is also of importance to those entering young adulthood, with approximately one third of individuals pursuing a degree higher than a bachelor's degree (Arnett, 2000).

Physical and Psychosocial Impacts of a Chronic Illness

Zhou and others (2016) looked at the medical records of 61 individuals with chronic illness to determine the impact of the emerging adulthood period of life both physically and psychosocially. As these individuals transitioned to adulthood, the efficacy of their transitions affected both their special health care needs and psychosocial development. These effects carried over into an individual's ability to consolidate identity, achieve independence and establish adult relationships – milestones that are typically reached during young adulthood (de Silva & Fishman, 2014).

When comparing individuals with and without a chronic illness, emerging adults with a chronic illness face lower success rates in finishing school, finding employment, and leaving their parental home. A meta-analysis further highlighted that individuals with a chronic illness experienced lower rates of marriage and becoming parents, and had lower income levels than healthy peers. The relationship between reaching developmental milestones within the emerging adulthood period of life and how well individuals with a chronic illness or disease fare is directly correlated (Pinquart, 2014).

Rationale for Present Study

To the best of our knowledge, no in-depth study of the daily life of emerging adults diagnosed with Ehlers Danlos syndrome has been conducted. The purpose of the present study was to gather more information about the experiences of individuals with classical and hypermobile EDS during the emerging adulthood period of life, specifically how these conditions impact social relationships during this dynamic developmental stage. The goal was to determine the impact, if any, that Ehlers Danlos syndrome has on individuals entering the emerging adulthood period of their lives, and to identify available resources for support. Our long term goal is that the data from this study will enable health care providers to better care for emerging adults with EDS.

METHODS

Participants

To capture the experiences of emerging adults with Ehlers Danlos Syndrome, we recruited participants between the ages of 20 and 30 years with a diagnosis of either hEDS or cEDS. Participants were recruited via snowball sampling; an email announcing the study was sent to the leader of the Ehlers-Danlos New England/Massachusetts Support Group and Chronic Pain Partners, who shared it with affiliated chapters nationwide. Recipients were further encouraged to share information about the study with other potential participants. Approximately three weeks after the initial recruitment email was sent, we followed up with a reminder email.

Instrumentation

We designed and hosted the anonymous survey using Qualtrics, an online survey software supported by Brandeis University. The survey consisted of 47 multiple choice, Likert scale and open-ended questions, which gathered both qualitative and quantitative data. Questions focused on categories including diagnosis, symptoms, relationships with family and friends, education, career planning, family planning and romantic partners, as well as demographic information. The Brandeis University Institutional Review Board determined that this study was exempt from full review.

Data Collection Procedure

An anonymous online survey was created through Qualtrics to collect our data. Participation was voluntary; individuals were free to skip any questions they did not wish to answer, or leave the survey at any time. Participants also had the option at the end of the survey to enter a raffle to receive one of four $25 Amazon.com gift cards and were directed to a separate, unlinked Qualtrics survey to enter their email address for the raffle. The survey was open from November 18, 2017 - December 25, 2017. Since participants were free to answer the questions they wanted there was variation in response rate from question to question.

Data Analysis

We analyzed our quantitative data for frequencies of responses as well as for associations and correlations using SPSS version 24. We analyzed the perceived impacts of Ehlers Danlos syndrome and its symptoms on relationships with family and friends, romantic relationships and family planning, education and career planning.

Responses to open-ended questions were manually analyzed by the lead author using an inductive approach to identify themes. The data from the open-ended survey questions were analyzed to determine themes that describe how EDS impacts the emerging adult period of an individual's life.

RESULTS

Survey Respondents

A total of 455 participants accessed the survey, of which 368 participants met eligibility criteria for participation in the study. As per enrollment criteria, respondents ranged in age from 20-30 years old, with a mean age of 25.0, and all carried a diagnosis of either cEDS or hEDS. Respondents were primarily female (90.9%) and lived across the United States as well as internationally (Table 1).

Table 1. Demographic Information		
EDS Type	Classical Hypermobile	40 (10.9%) 328 (89.1%) N=368
Gender	Female Male Other	291 (90.94%) 14 (4.38%) 15 (4.69%) N=320
Current Age	20-25 years 26-30 years	136 (47.9%) 148 (52.1%) N=284
Residency in the United States	Northeast Southeast Midwest Southwest West Outside the United States	88 (27.5%) 43 (13.44%) 71 (22.19%) 13 (4.06%) 32 (10%) 73 (22.81%) N=320
Highest Level of Education Level	Less than high school degree High school graduate or GED Some college but no degree Associates degree (2 year) Bachelor's degree (4 year) Graduate degree or higher (Master's, PhD, JD, MD, etc)	7 (2.21%) 25 (7.89%) 105 (33.12%) 28 (8.83%) 98 (30.91%) 54 (17.03%) N=317

Experience with Diagnosis of Ehlers Danlos Syndrome

We asked participants (n=368) when and how they were first diagnosed with EDS. A little over half of participants reported being diagnosed between the ages of 21 and 25 (54.78%). The greatest numbers of participants (31.37%) reported the EDS diagnosis was first brought up by a doctor based on their symptoms, while the second greatest number of participants (19.2%) first suspected the diagnosis based on an internet search of their symptoms.

The survey asked participants to select which of 11 different features of EDS they experienced. We combined these answers to determine that the five most common symptoms for those with EDS were joint hyper-flexibility (n=353), fatigue/fogginess (n=331), GI complications (n=304), musculoskeletal complications (n=298) and skin that bruises easily (n=276). The average number of symptoms was seven. We then asked participants to rank the severity of their EDS symptoms and calculated the mean severity rank for each symptom (Table 2). The five most severe symptoms reported by respondents were musculoskeletal complications, joint hyper-flexibility, fatigue/fogginess, Postural Orthostatic Tachycardia Syndrome (POTS), and GI complications (Table 2). The greatest number of participants (55.7%) described their EDS symptoms as somewhat severe.

Table 2. Most Common and Severe Symptoms Ranked

Most Common Symptoms	Frequency
1. Joint hyper-flexibility	354 (96.2%)
2. Fatigue/fogginess	332 (90.2%)
3. GI complications	305 (82.9%)
4. Musculoskeletal complications	299 (81.3%)
5. Skin that bruises easily	277 (75.3%)

Severity of Symptoms	Mean Severity
1. Musculoskeletal complications	3.47
2. Joint hyper-flexibility	3.59
3. Fatigue/fogginess	4.00
4. POTS (Postural orthostatic tachycardia syndrome)	4.13
5. GI complications	4.17
6. Skin that bruises easily	5.05
7. Fainting	5.35
8. Teeth/gum issues	5.39
9. Gynecological issues	5.53
10. Abnormal scarring	5.56

Table 2. The top five most common symptoms are ranked 1-5 from most to least common. The ranking of the severity of symptoms are ranked from most to least severe (1 being the most severe and 11 being the least severe).

Living with Ehlers Danlos Syndrome

A majority of participants (55.7%) described their EDS symptoms as somewhat severe. A little over half of participants (52%) described the physical as well as emotional impact of EDS as strongly negative (Table 3). Participants that noted their EDS symptoms as extremely severe also said that their social life with friends/peer relationships were greatly affected (87.5%) (Table 4). A majority of participants described the physical impacts of EDS as strongly negative (64.2%), while many (45.5%) also described EDS as having a mildly negative emotional impact on their life (Figure 1).

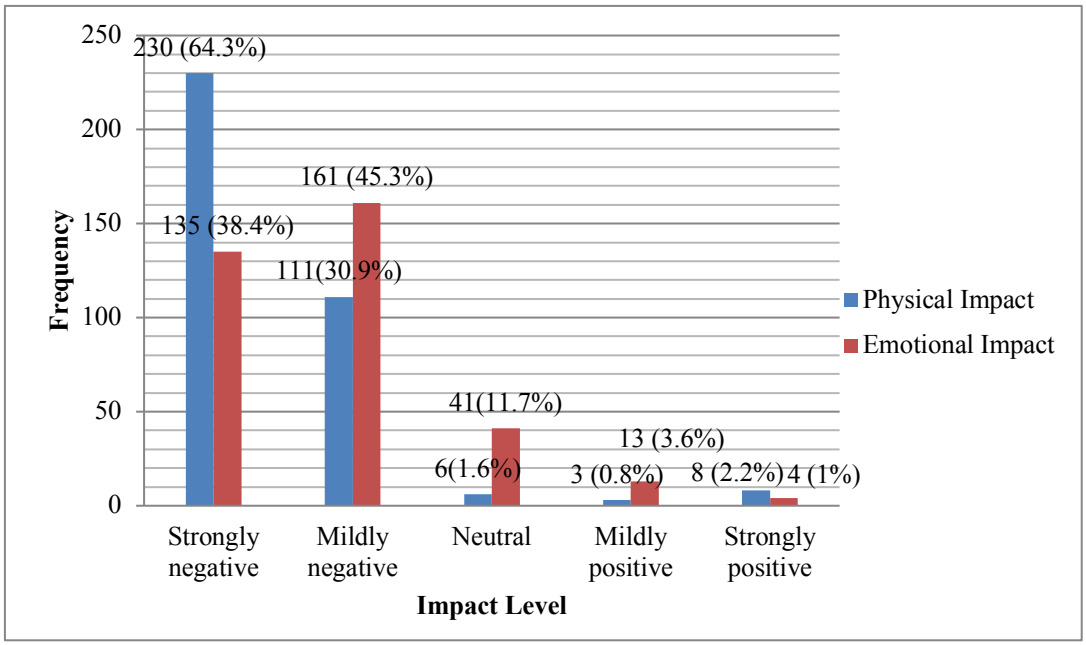

Figure 1. The level of physical and emotional impact that Ehlers Danlos syndrome has on survey respondents.

	Emotional Impact of EDS on One's Life	
Physical Impact of EDS on One's Life	Strongly negative	Somewhat negative
Strongly negative	119 (52%)	87 (38%)
Somewhat negative	15 (13.9%)	69 (63.9%)

Table 3. Directly comparing the emotional and physical effects of EDS and their impact on an individual's life. For example, for those participants who felt the physical impacts of EDS were strongly negative, 52% also felt that the emotional impact was strongly negative.

		How much would you say your diagnosis of EDS has impacted your social life with friends/peer relationships?					Total
		A great deal	A lot	A moderate amount	A little	None at all	
How would you describe the severity of your EDS symptoms?	Extremely severe	35	4	1	0	0	40
		87.5%	10.0%	2.5%	0.0%	0.0%	100.0%
	Somewhat severe	88	54	28	13	5	188
		46.8%	28.7%	14.9%	6.9%	2.7%	100.0%
	Neither mild nor severe	18	22	23	9	3	75
		24.0%	29.3%	30.7%	12.0%	4.0%	100.0%
	Somewhat mild	6	3	6	7	0	22
		27.3%	13.6%	27.3%	31.8%	0.0%	100.0%
	Extremely mild	0	1	0	0	1	2
		0.0%	50.0%	0.0%	0.0%	50.0%	100.0%

Table 4. A direct comparison between the severity of participants' symptoms and the impact level their diagnosis has had on their social life with friends/peers. Example: For those 87.5% of individuals who described their symptoms as extremely severe also reported that their social relationships were affected a great deal.

The greatest numbers of participants live with their significant other (35.1%), followed by living with their parents (27.8%). Similarly, when asked about their support system, the two most common were parents (31.38%) followed by significant others (24.69%) (Figure 2A). When attending medical appointments, about half of respondents attended these appointments by themselves (51.3%); others attended with their parent (31.7%) or significant other (11.24%) (Figure 2B). The average age of participants that attended doctor appointments with their parents was 23.3 years of age, while the average age of those who attended appointments with their significant other was 26.1 years of age. When the majority of a participant's support came from their patients, 46.6% of those participants went to the doctors with their parent(s).

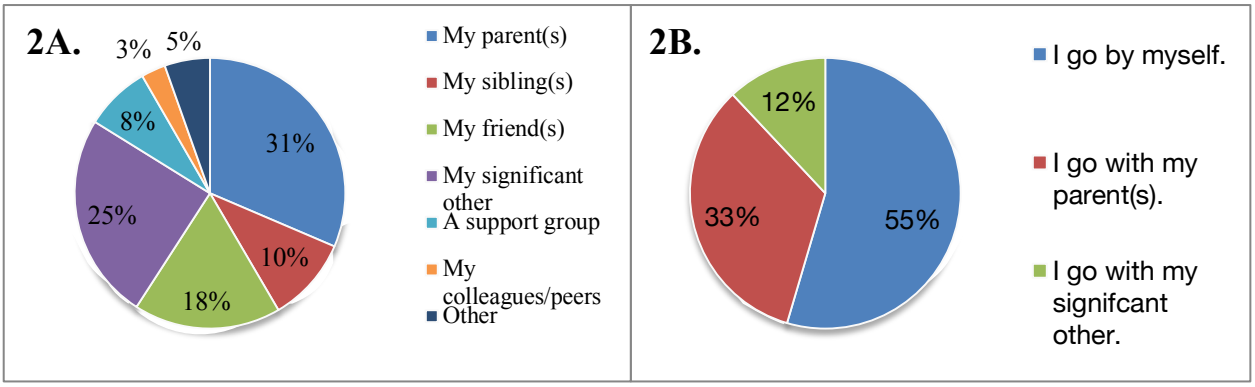

Figure 2A. The survey participants' support systems. Figure 2B. Attendees of medical appointments with the survey participants within the past two years.

Education and Career Planning

The education level of our participants ranged from less than a high school degree to an advanced degree, with most participants having completed or in the process of completing college (33.1%). Most individuals with EDS who responded to this study had at least a high school degree (Table 1).

The survey asked respondents to rank the impact that living with EDS had on their life choices. Participants strongly agreed that their diagnosis has impacted their ability to live independently (37.4%).The majority of respondents (40.5%) strongly agreed that their diagnosis

has prevented them from moving away for a job or school, and also influenced their career choice (54.0%). The greatest number of respondents (54.6%) feel that their diagnosis of EDS impacts their current career on a regular basis. A little over a quarter of participants work full time (>30 hours/week) (27.9%) and a third report not working (33.9%); receiving disability or applying for disability was a prevalent response among participants (18.2%).

Many participants offered advice to younger individuals who are thinking about moving away for school or a job:

"It's going to be hard. It's not going to be like anyone else's experience with EDS. Do your research and make sure to advocate for yourself or have someone who can do it for you… It's not worth it to push yourself beyond your limits and then pay for it later on a regular basis. Learn how to pace yourself and what energizes you. Get rid of as much as you can that takes your energy away." – 30 y.o. female diagnosed at age 6-10.

"Do it but be smart about it. I moved away to study. It's hard but I wouldn't change it for the world. Do move somewhere close to medical teams. Do embrace independence of however it may look to you … If the place of study doesn't have a solid supportive disability inclusion team and you know you have troubles with sickness, dislocations, etc then don't go. Go to a place of study that will cater to your needs. While there will be times you will feel isolated and alone, connect with someone… Goals and dreams, passions and desires – these are protective measures. These are the things that will get you out of bed on a crap morning." – 27 y.o. female diagnosed at age 21-25.

Relationships

Participants were either single (40.5%), in a committed relationship (29.9%), or married (27.4%). We asked them to rate the level of impact they feel EDS has had on six different aspects of their romantic relationships. Most participants felt that EDS impacted all six aspects, particularly sexual intimacy (3.44). Participants felt that they were secondly most impacted in "allowing yourself to be vulnerable with someone" (mean 3.3) followed by "maintaining relationships" (mean 3.1). Single individuals experienced a higher impact in all categories that were looked at, but this data was not statistically significant. Those who were married experienced less impact in all categories except sexual intimacy (Figure 3).

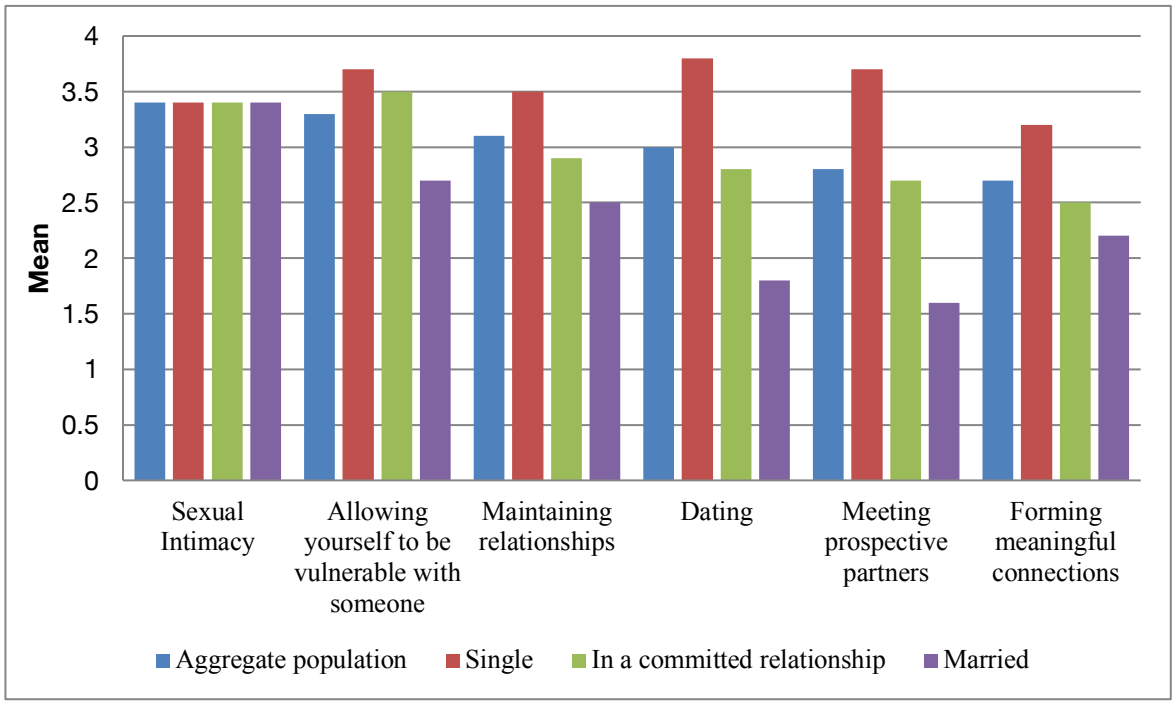

Figure 3. Six aspects of romantic relationships were studied, with sexual intimacy overall being most highly affected by those with EDS. The higher the mean, the more severely each area is impacted in romantic relationships.

There was a significant correlation between the severity of an individual's EDS symptoms and their perception that their relationships were impacted. The more severe an individual's symptoms were, the greater the impact they felt on their romantic, family member, and peer relationships. Furthermore, there was a significant correlation between the number of symptoms an individual reported and romantic relationships, family member relationships, and friendships; the more affected the participant was, the greater the impact on these three groups. Current age was significant for romantic relationships and family member relationships. The older a participant was, the more significant their age was in impacting relationships with partners and family (Table 5).

	Romantic relationships	Family relationships	Friendships
Severity of symptoms	0.408 *** Std err: 0.076	0.245 ***	0.198 ** Std err: 0.060
Number of symptoms	0.140 *** Std err: 0.030	0.065 **	0.085 *** Std err: 0.023
Current age	-0.049 ** Std err: 0.016	-0.026 **	-0.012 Std err: 0.013
R^2	0.228	0.135	0.113

Table 5. A regression analysis between romantic relationships, family relationships and friendships was conducted. These were compared to the severity of an individual's symptoms, the number of symptoms, and the participant's current age. N = *** sig @ $p<0.001$; ** sig @ $p<0.05$.

We went on to ask participants about their experiences with having a family history of EDS or being the first/only person diagnosed. The vast majority of respondents (81.25%) reported that they were the first person in their family to be diagnosed with Ehlers Danlos Syndrome. Of those diagnosed, 20.2% of participants were the only affected individual in their family. Just over 32% of participants reported having another formally diagnosed family member, and nearly half (47.4%) had family member(s) with suspected EDS. Open-ended questions about being the first individual diagnosed with EDS versus having a family member already diagnosed with EDS revealed themes regarding isolation, feeling misunderstood, and (for those with previously-diagnosed family members), mixed feelings. Some felt as if they were in constant comparison with their family members to determine whose symptoms were more severe, while others felt a sense of validation and alliance with their family.

Several individuals that were the first or only person in their family to be diagnosed with EDS mentioned feeling misunderstood or isolated:

"My family thinks that i'm [sic] lying. They will never understand" – 30 y.o. female diagnosed at age 16-20.

"…I isolate myself from other [sic] as to not burden them. I often keep the truth of what I am going through everyday from my parents so they are not aware of how much suffering I go through so as to not add more pain to their lives for no reason." – 21 y.o. female diagnosed at age 21-25.

Some participants with a family history alluded to the support and validation that came from having a family member who understands:

"It is helpful that my dad also experiences some similar struggles as I do. He understands what it's like to have chronic pain and he just gets it more than other people who don't have it." – 24 y.o. female diagnosed at age 11-15.

"I believe it has affected my personal experience in a positive way for the following reasons: 1) I had someone to talk to who knows what I'm going through 2) I probably wouldn't have been diagnosed because I didn't know it existed outside my mother's experience 3) we help each other navigate ways to manage symptoms" 28 y.o. female diagnosed at age 21-25.

However, other respondents with a family history of EDS reported a much less positive experience, mentioning an underlying feeling of competition surrounding their symptoms:

"My sisters symptoms were always much more severe so I always felt like mine were measured to that." – 30 y.o. female diagnosed at age 0-5.

"My grandmother and mom also have eds and I struggle sometimes because my grandmother is constantly trying to compare her and my pain, she wants me to tell her that I'm not hurting as much as she is, but I feel like everyone is just trying to survive this life and who can say that one agony is equal to another." – 21 y.o. female diagnosed at age 16-20.

In describing their relationships, many participants noted feelings of embarrassment or anxiety about their physical needs and worries about rejection from romantic partners and friends. However, many also noted that romantic partners, family, and friends have been supportive and understanding despite these concerns.

On romantic relationships:

"I worry about the burden I am on my fiancé. It is a lot for him to take on. He is aware of the potential future I have and the disabling portions of my disease. My mom and I have given him many opportunities to walk away and he always stays. I could not imagine getting through every day without his support." – Female diagnosed at age 16-20.

"The physical limitations caused by EDS limit my ability to participate in activities that are common among my peers in a dating scenario [sic] (going out at night, drinking alcohol, any physically strenuous activities). It is difficult to connect with potential partners as medical treatment and issues take up the majority of my time and energy." – Female diagnosed at age 21-25.

"We have difficulties with physical intimacy because I am in pain 24/7 365 and he doesn't initiate anymore because he doesn't want to hurt me. I know this is out of love, but sometimes I feel like he doesn't find me attractive anymore." – 29 y.o. female diagnosed at age 21-25.

"I'm unable to have sex anymore due to the pain from EDS." – 30 y.o. female diagnosed at age 6-10.

On friendships:

“I don’t feel like I can connect with them as much anymore. There’s too much they can’t understand. It makes me feel alone around them. I miss out on a lot of things I’m invited to which creates distance with friends. I’m embarrassed of some of my symptoms which makes me want to stay home even more when I’m feeling unwell.” – 27 y.o. female diagnosed at age 21-25.

“I’ve lost friend [sic] because they don’t understand the condition or why I may cancel last minute. I have a great support group of friends who empathize with me. My circle isn’t huge but they are amazing people.” – 27 y.o. female diagnosed at age 21-25.

“I was stunned by the great lengths my friends take to support me like always make sure that I have somewhere to sit whenever we go out. It took a lot of therapy before I could open up and articulate my needs but once I did my friends went above and beyond.” – 25 y.o. female diagnosed at age 21-25.

DISCUSSION

Through this survey, we were able to collect data enabling us to better characterize how being diagnosed with Ehlers Danlos syndrome has impacted the lives and decision making of emerging adults. We focused particularly on some of the less tangible aspects of relationship forming, and areas of development that are often important to emerging adults: education, career planning, and social relationships. A third of individuals were receiving disability benefits. A third of individuals felt that their diagnosis impacted their career choice; however, the highest education level attained was similar to that of typical emerging adults. Relationships with romantic partners, family members and friendships were significantly impacted. The diagnosis had a significant physical and emotional impact on individuals. Our goals were to characterize the unique challenges for emerging adults with EDS and to highlight the needs of this group so they can be most appropriately supported by their healthcare providers throughout their lives.

Diagnosis

Joint hypermobility and fatigue were the two most common symptoms of EDS reported by our participants. Voermans et al. conducted a study with a Dutch EDS registry and concluded that chronic pain is highly prevalent and more severe in the hypermobility type than classic type. Pain is also associated with hypermobility, dislocations and surgery, all which contribute to functional impairment in daily life and fatigue severity (Voermans et al., 2010). For the purpose of our study, we focused on the most common EDS types, and did not subdivide our participants

by diagnosis. The results of our study align with previous research in terms of the most limiting and common symptoms of EDS.

Education and Career Planning

Although none of the variables we analyzed were found to have a statistically significant impact on education or career, the variability in participant responses and the responses to the open-ended questions indicate that they may have a significant impact on an individual basis. According to Pinquart et al. (2014), individuals with a chronic illness have lower rates of success finding employment and leaving the parental home, just as our participants highlighted that they were not able to move away for school or a career opportunity because of their EDS.

When comparing the data of the study population to the 2015 Education Attainment Assessment (Educational Assessment in the United States: 2015), which provides a portrait of educational attainment from the Current Population Survey (CPS), respondents with EDS and their typical peers had similar levels of educational completion. Although participants mention that they feel as if their condition has an impact on their life, the data shows they may not be that different as far as educational achievements (Table 6).

	EDS Individuals	**"Typical" Individuals**
High school graduate or more	97.8%	90.5%
Some college or more	64.7%	65%
Associate's degree or more	55.8%	46.5%
Bachelor's degree or more	24.9%	31.6%
Advanced degree	7.9%	10.9%

Table 6. Educational Attainment of EDS respondents and their peers. (Of note the 2015 Education Attainment Assessment is using an age range of 25-34 years of age while the EDS population studied is 20-30 years of age.)

Relationships

Relationships, whether romantic, family, or friendship oriented, were significantly impacted for respondents with EDS who completed the survey. Living with a chronic condition that is life-altering can leave someone with EDS feeling misunderstood and burdensome to those around them. There is a negative significant correlation between both severity of symptoms and number of symptoms, and relationships with friends, family, and partners. Relationships were measured by comparing the severity of symptoms, number of symptoms, and a participant's age (Table 5). This supports the reviewed literature that having a different identity than others their age affects someone's ability to achieve independence and establish long lasting, adult relationships (de Silva & Fishman, 2014).

Romantic relationships during emerging adulthood are typically temporary, as an individual learns what they need from a mate. Therefore, the variability in responses regarding romantic relationships may be a result of having EDS, but may also be a result of emerging adulthood in general. During this time, emerging adults are changing their allegiance from parents to romantic partners, but it is more challenging for those with EDS. Individuals who were not in a relationship experienced higher impact on the six aspects of romantic relationships that were studied. Many individuals responded that sexual intimacy was the most impacted aspect of their romantic relationships, with open-ended responses touching upon the pain that they experience during sexual activities, as well as fatigue that sometimes does not allow for any intimacy at all. These results are consistent with the significant impact of EDS on romantic relationships, with both the number and severity of symptoms playing a role.

Additionally, even respondents who were married or in a committed relationship and felt supported by their partner worried that their significant other would leave them due to all of the care they would require moving forward. Our participants reported that even supportive partners never really understood their diagnosis of EDS, leaving them in a state of isolation. They reported stress from not being able to equally contribute to household chores, as well as about partners becoming caregivers. Some participants worried about losing independence and becoming burdensome in a new marriage, taking an immense emotional toll on both individuals.

Another area of romantic relationships that was greatly impacted was "allowing yourself to be vulnerable with someone." Many respondents were concerned that their diagnosis would scare away potential partners. They did not want to open up about the difficulties they experienced, or care and lifestyle modifications they may require. Furthermore, respondents reported difficulties connecting with other peers their age and meeting prospective partners because they could not fully immerse themselves in typical activities, and worry that prospective partners would not accept them for who they are.

Participants had mixed feelings when discussing family relationships. An individual's severity of symptoms, number of symptoms and current age played a significant role in family relationships. While some individuals consistently received support from their family members when receiving a diagnosis of EDS, others did not have a familial support system that they could rely upon. In some instances, individuals who had a family history of EDS felt as if family members were comparing their symptoms to one another in an effort to make one feel lesser than the other, while other family members were not able to come to terms with an invisible condition, a condition that cannot be seen, much less the diagnosis of EDS and the symptoms that participants were facing.

Friendships were significantly impacted based on a participant's severity of symptoms and number of symptoms. Open-ended responses regarding friendships were generally negative in nature, indicating that having a chronic condition impacts social aspects of an individual's life. Living with EDS also made friendships difficult due to the discomfort of conversations about the condition and having to explain why someone will not be able to get together due to fatigue or other physical limitations. Others felt deserted and isolated. Some respondents had many friends, but as the symptoms of EDS began to worsen, only a few remained. However, other open-ended responses consisted of an expression of gratitude to their close friends who did understand and did not leave their sides when they needed them the most.

Overall, medical professionals need to be aware of emerging adults with Ehlers Danlos and the types of impacts the condition has on relationships with family, friends, and romantic partners. During emerging adulthood, individuals transition from relying on parents to relying on peers. If relationships with friends or significant others is not strong, emerging adults with EDS may have fewer emotional supports than individuals in other age groups. Individuals with EDS may face a greater emotional burden of having a diagnosis, and health care professionals should be cognizant of the patient's available support system.

Living with Ehlers Danlos Syndrome

Our study revealed a strong relationship between the reported physical and emotional impacts of EDS on respondents. Participants who reported extremely severe or somewhat severe symptoms were more likely to report that EDS had a negative physical and emotional impact on their lives. This supports previous research, which found that living a restricted life with EDS can cause fear, pain, and stigmatization, further impacting emotional and physical responses

(Berglund et al., 2000). A study performed with almost 300 individuals with EDS assessed fatigue severity, functional impairment in daily life, physical activity, psychological distress, social functioning and more; results indicated that more than three quarters of EDS patients suffer from severe fatigue. These patients present with a higher level of psychological distress, showing that fatigue is a clinically relevant problem in EDS (Voermans et al., 2010).

Furthermore, respondents' most limiting factors included musculoskeletal complications and joint hyper flexibility which caused pain, as well as fatigue, hindering them from interactions with peers and other aspects of adulthood. Individuals with more limiting physical symptoms are more likely to withdraw themselves from society and therefore, we can infer that those with more limiting physical symptoms have a more difficult time transitioning to adulthood. They are more likely to rely on their family more so than those with less limiting physical symptoms. A strong negative physical impact can lead to a negative emotional impact on an individual. Our study demonstrates that this hinders an individual's transition to adulthood, as well as a further dependence on parental support.

A majority of our participants reported that their support system came primarily from their parent(s), highlighting their inimitable support during a challenging period of time. A third of participants continue to attend doctor's appointments with their parents within the last two years, although older participants were more likely than younger participants to attend appointments with a significant other. Underlying medical issues have the ability to affect different relationships, and having the continuous support of family compared to a romantic partner may feel more comfortable for those with EDS.

An individual coming to clinic with their parent as opposed to by themselves or with a significant other may be in a different place either in diagnosis or life, and medical professionals may want to take note this information when meeting a new patient. Also, while physical symptoms may be the only focus of the appointment, healthcare providers should also be aware of the emotional needs and the potential lack of a support network in this age group.

Practice Implications

Through this study we have the ability to impact the clinical care for those with EDS. Many individuals in this study expressed their concerns that medical professionals do not understand their condition and they feel misunderstood, misguided, and dismissed in a time of need. Patients reported that, although their condition is invisible, their pain is real and they wish that their doctors and medical professionals would really listen to and try to understand their issues.

As genetic counselors, although we cannot solve the patient's symptoms, we can listen and validate their concerns. Genetic counselors that are involved with the diagnosis of an individual with EDS during this time period should be aware of the social challenges. Patient support can be offered, as well as acknowledging the challenges that can arise with peers, romantic partners, and family members. Patients may also find it helpful to role play a scenario on how to deliver information about their diagnosis or their needs to important people in their life. Although there is less research regarding direct role play with patients and its effectiveness, research has demonstrated that role-play was valued by medical students in the acquisition of communication skills even though some had prior unhelpful experiences (Nestel & Tierney, 2007).

Referrals can be made to support groups, physical and occupational therapy, and pain management programs. Although a genetic counselor may be able to provide initial counseling to the patient, a referral to a psychologist or support group may benefit some patients who wish to delve more deeply into concerns relating to different aspects of life during emerging adulthood and coping mechanisms. Genetic counselors as health care professionals have the ability to directly impact these individuals' lives and taking these steps can improve their quality of life and their transition to emerging adulthood.

LIMITATIONS

Those participants who chose to opt-in to our study or answered our open-ended responses may have stronger or different opinions than those individuals with EDS who did not. Our survey also had the inability to tease out differences based on symptoms given the survey design as these were "select all" questions. Furthermore, we recruited participants through an email that was sent by one person to support groups across the United States and internationally, and therefore our results reflect those individuals who are a part of a support group. Different parts of the United States were well represented, but recruiting through other venues may have reached additional people with EDS. Finally, our demographics were heavily skewed towards female participants and those with hEDS.

RESEARCH RECOMMENDATIONS

Given the exploratory nature of this research, much of the measures of psychosocial functioning were based on analysis of open-ended, qualitative questions to identify common themes and allow for respondents to more openly share their experiences. Based on the negative impact of emotional and physical limitations symptoms have on individuals with EDS, future research could be utilized to assess general measures of these impacts across this age group as well as the impact on social relationships. Potential support groups specifically for young and/or emerging adults could be targeted utilizing a validated tool to further investigate coping mechanisms for social isolation. Interviews could be an avenue for further study to explore specific social challenges of those with EDS more in depth.

Additionally, a prevalent theme throughout the responses was a feeling that health care providers were not well informed about Ehlers Danlos syndrome. Much of this could be due to the variability of the symptoms, but further research could look into this patient provider disconnect in more detail or develop additional resources for health care professionals to share with their patients.

CONCLUSION

Although emerging adulthood is characterized as a time of exploration, change, and new beginnings, having a chronic condition such as Ehlers Danlos syndrome impacts this period of transition. Our results suggest that romantic relationships, family relationships and social relationships are all significantly negatively impacted. While many individuals demonstrated resiliency and the ability to transition, others struggled with the diagnosis. Furthermore, medical professionals have the ability to positively impact their patients by utilizing listening skills. Medical professionals can also provide support by working with their patient to come up with treatment that is individualized to their specific needs.

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APPENDICES

APPENDIX A: RECRUITMENT NOTICE

Hello! My name is Katie Bealka, and I am a graduate student in the Brandeis University Genetic Counseling Master's degree program. I am looking to better what life is like for emerging adults living with Ehlers Danlos Syndrome (EDS), either the classical or hypermobile type, and need your help.

Have you been diagnosed with either the classical or hypermobile type of Ehlers Danlos syndrome?

Are you currently between the ages of 20 and 30 years old?

If so, we invite you to participate in our anonymous, online survey. *The survey will ask questions about your experiences living with Ehlers Danlos syndrome as a young adult and should take approximately 10-15 minutes of your time.* Your responses will be kept anonymous, and as a token of appreciation for participating, you'll have the option to enter a raffle to win one of three $25 gift cards to **Amazon.com**. We hope that the results of this survey will help educate medical professionals about the unique needs of young individuals with EDS.

This study has been approved by the Brandeis University Institutional Review Board.

CLICK HERE: for more information or to participate in the survey.

If you have any questions about this research project please contact Katie Bealka at **kbealka@brandeis.edu**. If you have questions about your rights as a research participant please contact the Brandeis Institutional Review Board at 781- 736-8133 or **irb@brandeis.edu**.

We'd also love if you share this study with any family members and/or friends that you think might qualify, as we'd love to get a large response.

EMAIL CONSENT

Hi Katie,

I would be happy to help you circulate your EDS Study to my groups and many contacts I have around the U.S. and the World. When you have it ready, please write an introduction that includes how people who participate will learn the results of the study. With so many surveys/studies going on (which is great) people like to understand how their information will be used and what kind of feedback they will receive once the study is done. Also, the possibility of it helping to instigate further research. Our group has helped initiate several studies (including IRBs) and are promoting or participating in many others.

Always feel free to contact me with questions or to talk,

My Best,
Jon

p.s. I have provided my full contact information for you and your students/colleagues to have.

Jonathan Rodis

President-Massachusetts Chapter of the Marfan Foundation
Founder & Co-chair-Ehlers-Danlos New England/Massachusetts Support Group
Founder & Chair-Physician Awareness Committee(s) for Marfan & Ehlers-Danlos Syndrome(s)
National Disability & Medical Advocate for Rare Disorders
Member: Winthrop Disability Commission
Member- Ambry Genetics Advisory Board
Member- Dysautonomia Support Network Board
CTDAwareness2013 on Inspire.com
Living With Marfan Syndrome: www.jrmarfan58.com

APPENDIX B: STUDY INSTRUMENT

Ehlers Danlos Syndrome: Navigating the Transition through Emerging Adulthood

Q1 Hello,

You are invited to take part in this study because you are within the ages of 20 and 30 years old, and have been diagnosed with either hypermobile or classic Ehlers Danlos syndrome.

If you agree to take part in this study, it should take you approximately 10 minutes to complete the survey. At the end of the survey, you will be directed to a separate page where you have the opportunity to enter your email address into a drawing for one of four $25 Amazon.com gift cards. Your email address will not be linked to your survey responses, and will be deleted once the gift card winners have been notified.

There is no direct benefit to you for taking part in this study. Participation in this study will help us to understand more about the impact of Ehlers Danlos on the emerging adulthood period of life. There are no known risks to take part in this study.

The information collected within the study will not contain identifying information. You may choose not to participate in this study and can skip any questions that make you uncomfortable. You may also exit the study and withdraw at any time.

If you have any questions about this research project please contact Katie Bealka at kbealka@brandeis.edu, or the Brandeis University faculty sponsor, Cassandra Buck, at cbuck@brandeis.edu.

We appreciate you taking the time to complete this survey. If you have questions about your rights as a research participant please contact the Brandeis Institutional Review Board at 781-736-8133 or irb@brandeis.edu.

If you are eligible and agree to take part in the study, click the 'Continue to survey' button below.

- ○ Continue to survey (1)
- ○ End survey (2)

Skip To: End of Survey If Hello, You are invited to take part in this study because you are within the ages of 20 and 30 ye... = End survey

Q2 **Inclusion Criteria**
Are you currently between the ages of 20 and 30?

- ○ Yes (1)
- ○ No (2)

Skip To: End of Survey If Inclusion Criteria Are you currently between the ages of 20 and 30? = No

Q3 Have you been diagnosed with Classical or Hypermobile Ehlers Danlos syndrome?

- ○ Yes, Classical EDS (1)
- ○ Yes, Hypermobile EDS (2)
- ○ No (3)

Skip To: End of Survey If Have you been diagnosed with Classical or Hypermobile Ehlers Danlos syndrome? = No

Q4 **EDS Experience:** The following questions will help us to better understand your experience with Ehlers Danlos syndrome and how you were diagnosed.

How old were you when you were first diagnosed with Ehlers Danlos syndrome?

- ○ 0-5 (1)
- ○ 6-10 (2)
- ○ 11-15 (3)
- ○ 16-20 (4)
- ○ 21-25 (5)
- ○ 26-30 (6)

Q5 How did you first learn you might have EDS (select all that apply)?

- ☐ Internet search of my symptoms (1)
- ☐ Clinical examination by a doctor (2)
- ☐ A physical therapist (3)
- ☐ A rheumatologist (4)
- ☐ A friend suggested it (5)
- ☐ A family member suggested it (6)
- ☐ A personal trainer (7)
- ☐ Other, please describe (8) _____

Q6 Did you have genetic testing for Ehlers Danlos syndrome?

- ○ Yes (1)
- ○ No (2)

Display This Question:

If Did you have genetic testing for Ehlers Danlos syndrome? = Yes

Q7 What was the result of this test?

- ○ Positive (EDS-causative variant identified) (1)
- ○ Negative (EDS variant not identified) (2)
- ○ Inconclusive result (variant of uncertain significance, VUS) (3)
- ○ I do not remember (4)

Q8 How would you describe the physical impact of EDS on your life?

- ○ Strongly negative (1)
- ○ Mildly negative (2)
- ○ Neutral (3)
- ○ Mildly positive (4)
- ○ Strongly positive (5)

Q9 How would you describe the emotional impact of EDS on your life?

- ○ Strongly negative (1)
- ○ Mildly negative (2)
- ○ Neutral (3)
- ○ Mildly positive (4)
- ○ Strongly positive (5)

Q10 Please tell us more about your specific EDS symptoms. Which of the following features do you have (select all that apply)?

- [ ] Joint hyperflexibility (1)
- [ ] Musculoskeletal complications (2)
- [ ] Skin that easily bruises and/or scars (3)
- [ ] GI complications (pain, bloating, constipation, diarrhea, etc) (4)
- [ ] Gynecological issues (5)
- [ ] Teeth/gum issues (6)
- [ ] POTS (Postural orthostatic tachycardia syndrome) (7)
- [ ] Fatigue/fogginess (8)
- [ ] Other: (9) _____

Q11 How would you describe the severity of your EDS symptoms?

- ○ Extremely severe (1)
- ○ Somewhat severe (2)
- ○ Neither mild nor severe (3)
- ○ Somewhat mild (4)
- ○ Extremely mild (5)

Carry Forward Selected Choices from "Please tell us more about your specific EDS symptoms. Which of the following features do you have (select all that apply)?"

Q12 Rank your symptoms of EDS from least (1) to most severe (up to 9) based on the impact they have on your life (drag to reorder symptoms):

_____ Joint hyperflexibility (1)
_____ Musculoskeletal complications (2)
_____ Skin that easily bruises and/or scars (3)
_____ GI complications (pain, bloating, constipation, diarrhea, etc) (4)
_____ Gynecological issues (5)
_____ Teeth/gum issues (6)
_____ POTS (Postural orthostatic tachycardia syndrome) (7)
_____ Fatigue/fogginess (8)
_____ Other: (9)

Q13 **Personal and Family History:**
Are other members of your family affected with Ehlers Danlos syndrome?

- ○ Yes (1)
- ○ No (2)
- ○ Not clinically diagnosed, but he/she/I believe he/she also has EDS (3)

Q14 Who was the first person in your family to be diagnosed with Ehlers Danlos?

- ○ I am the first person in my family. (1)
- ○ My mother/father was the first person. (2)
- ○ My brother/sister was the first person. (3)
- ○ Another relative was the first person. Please specify (4)

_____

Display This Question:

If Personal and Family History: Are other members of your family affected with Ehlers Danlos syndrome? = Yes

Q15 Do you feel like having a family history of EDS has affected your personal experience with the syndrome? Please elaborate:

_____

_____

_____

_____

_____

Q16 **Relationships:**

Do you feel like living with Ehlers Danlos syndrome has affected your relationship with:

	Yes (1)	No (2)
Your parents? (1)	☐	☐
Your siblings? (2)	☐	☐
Your peers/colleagues? (3)	☐	☐
Romantic partners? (4)	☐	☐

Q17 What is your current relationship status?

- ○ Single (1)
- ○ In a committed relationship (2)
- ○ Married (3)
- ○ Domestic partnership (4)
- ○ Separated (5)
- ○ Divorced (6)
- ○ Widow/Widower (7)

Q18 Who do you live with currently (select all that apply)?

- ☐ With parent(s) (1)
- ☐ With sibling(s) (2)
- ☐ With friend(s) (3)
- ☐ With significant other (4)
- ☐ By yourself (5)
- ☐ Other: (6) _____

Q19 Where does the majority of your support system come from (select all that apply)?

- [ ] My parent(s) (1)
- [ ] My sibling(s) (2)
- [ ] My friend(s) (3)
- [ ] My significant other (4)
- [ ] A support group (5)
- [ ] My colleagues/peers (6)
- [ ] Other: (7) _____

Q20 **Within the last 2 years**, who typically accompanies you to your doctors' appointments (select all that apply)?

- ○ I go by myself. (1)
- ○ I go with my parent(s). (2)
- ○ I go with my sibling(s). (3)
- ○ I go with my friend(s). (4)
- ○ I go with my significant other. (5)
- ○ Other: (6) _____

Q21 How much would you say your diagnosis of EDS has impacted the following aspects of your **romantic relationships**? Please rate the following statements.

	A great deal (1)	A lot (2)	A moderate amount (3)	A little (4)	None at all (5)
Meeting prospective partners (1)	○	○	○	○	○
Forming meaningful connections (2)	○	○	○	○	○
Dating (3)	○	○	○	○	○
Allowing yourself to be vulnerable with someone (4)	○	○	○	○	○
Sexual intimacy (5)	○	○	○	○	○
Maintaining relationships (6)	○	○	○	○	○

Q22 Please use the space below to elaborate on any of the above aspects or tell us more about how your diagnosis of EDS has or has not impacted your **romantic relationships.**

_____

_____

_____

_____

_____

Q23 How much would you say your diagnosis of EDS has impacted the following aspects of your relationship with **family members (immediate or extended)**? Please rate the following statements.
Living with Ehlers Danlos syndrome has:

	A great deal (1)	A lot (2)	A moderate amount (3)	A little (4)	None at all (5)
Helped me know who I can count on in times of trouble. (1)	○	○	○	○	○
Makes me more willing to help others. (2)	○	○	○	○	○
Helped relationships become more meaningful. (3)	○	○	○	○	○
Helped me become closer to people I care about. (4)	○	○	○	○	○
Helped me become more aware of the love and support available from other people. (5)	○	○	○	○	○

Q24 Please use the space below to elaborate on any of the above aspects or tell us more about how your diagnosis of EDS has or has not impacted your relationship with **family members.**

_____

_____

_____

_____

_____

Q25 How much would you say your diagnosis of EDS has impacted the following aspects of your relationship with **friends**? Please rate the following statements.

	A great deal (1)	A lot (2)	A moderate amount (3)	A little (4)	None at all (5)
Helped me know who I can count on in times of trouble. (1)	○	○	○	○	○
Makes me more willing to help others. (2)	○	○	○	○	○
Helped relationships become more meaningful. (3)	○	○	○	○	○
Helped me become closer to people I care about. (4)	○	○	○	○	○
Helped me become more aware of the love and support from other people. (5)	○	○	○	○	○

Q26 Please use the space below to elaborate on any of the above aspects or tell us more about how your diagnosis of EDS has or has not impacted your relationship with **friends**.

_____

_____

_____

_____

_____

Q27 How much would you say your diagnosis of EDS has impacted your social life with friends/peer relationships?

- ○ A great deal (1)
- ○ A lot (2)
- ○ A moderate amount (3)
- ○ A little (4)
- ○ None at all (5)

Q28 Please use the space below to elaborate on any of the above aspects or tell us more about how your diagnosis of EDS has or has not impacted your **social like with friends/peers.**

_____

_____

_____

_____

_____

Q29 **Family Planning:**
Do you have children?

- ○ Yes, biological (1)
- ○ Yes, adopted (2)
- ○ No, I do not have children (3)

Display This Question:
If Family Planning: Do you have children? = Yes, biological

Q30 Did you have your children before you were diagnosed with EDS?

- ○ Yes (1)
- ○ No (2)

Display This Question:
If Family Planning: Do you have children? = Yes, biological

Q31 Do you plan to have more children?

- ○ Yes (1)
- ○ Maybe (2)
- ○ No (3)

Display This Question:
If Family Planning: Do you have children? = Yes, biological

Q33 To what extent has your diagnosis of EDS influenced this position?

- A great deal (1)
- A lot (2)
- A moderate amount (3)
- A little (4)
- None at all (5)

Display This Question:

If Family Planning: Do you have children? = Yes, adopted

Q34 Did your diagnosis of EDS play a role in adopting children?

- Yes (1)
- No (2)

Display This Question:

If Did your diagnosis of EDS play a role in adopting children? = Yes

Q35 How so?

_____

_____

_____

_____

_____

Display This Question:

If Family Planning: Do you have children? = No, I do not have children

Q36 Do you plan to have children?

- ○ Yes (1)
- ○ No (2)
- ○ Undecided (3)
- ○ Haven't thought about it (4)

Display This Question:

If Family Planning: Do you have children? = No, I do not have children

Q37 To what extent has your diagnosis of EDS influenced this position?

- ○ A great deal (1)
- ○ A lot (2)
- ○ A moderate amount (3)
- ○ A little (4)
- ○ None at all (5)

Q38 If you would like to tell us more about how your diagnosis has affected your decisions to have or not have children, please use this space below.

_____

_____

_____

_____

_____

Q39 **Career Planning/Exploration**
Please rate your agreement with the following statements.

	Strongly agree (1)	Agree (2)	Somewhat agree (3)	Neither agree nor disagree (4)	Somewhat disagree (5)	Disagree (6)	Strongly disagree (7)
My diagnosis limits my ability to move away for school or a job. (1)	○	○	○	○	○	○	○
My diagnosis has had an influence on my career choice. (2)	○	○	○	○	○	○	○
My diagnosis has impacted my ability to live independently. (3)	○	○	○	○	○	○	○

Q40 Do you feel that your diagnosis of EDS impacts your current career on a regular basis?

- ○ Yes (1)
- ○ No (2)
- ○ I am not working right now. (3)
- ○ I do not know. (4)

Q41 **General:**
How can medical professionals (doctors, genetic counselors, surgeons) better serve your needs with this diagnosis?

_____

_____

_____

_____

_____

Q42 What advice would you give someone who is newly diagnosed with EDS about life living with the condition?

_____

_____

_____

_____

_____

Q43 What advice would you give to a young adult with EDS who is thinking about going to college or moving away from home?

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Q44 Thinking back to when you were 18 years old, what advice would you give yourself, or do you wish someone had given you?

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_____

Q45 **Final Demographics**: Please tell us a little bit more about you.
What is your current age in years?

_____

Q46 What is your gender?

- ○ Male (1)
- ○ Female (2)
- ○ Other, I describe myself as (3) _____

Q47 In what part of the United States do you reside?

- Northeast (1)
- Southeast (2)
- Midwest (3)
- Southwest (4)
- West (5)
- Outside of the United States (6) _____

Q48 What is your highest level of education?

- Less than high school degree (1)
- High school graduate or GED (2)
- Some college but no degree (3)
- Associates degree (2 year) (4)
- Bachelor's degree (4 year) (5)
- Graduate degree or higher (Master's, PhD, JD, MD, etc) (6)

Q49 What is your current occupation (select all that apply)?

- ☐ I work full time (>30 hours/week). (1)
- ☐ I work part time ((2)
- ☐ I am a full time student. (3)
- ☐ I am a part time student. (4)
- ☐ I am receiving disability. (5)
- ☐ Other: (6) _____

Q50 *Thank you for your thoughtful responses and time spent taking my survey. Your responses have been saved. If you would like the chance to win one of four $25 Amazon gift cards, please click the following link. You will be directed to a separate survey, unlinked to this data, where you can enter your email address for the gift card raffle.*

- ○ 'Following link that will be added' (1)
- ○ No than you, end survey. (2)

Skip To: End of Survey If Thank you for your thoughtful responses and time spent taking my survey. Your responses have been... = No than you, end survey.

End of Block: Default Question Block