Understanding the psychosocial impact of Joint Hypermobility Syndrome and Ehlers-Danlos Syndrome Hypermobility Type: A qualitative interview study

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Declaration of interest

The authors report no declarations of interest.

Implications for rehabilitation

- Participants who had help from family members to complete activities described guilt and shame, highlighting the need for a greater rehabilitation focus on maintaining independence.
- Difficulties with sexual relationships due to prolapse or erectile dysfunction, and associated anxieties have indicated a need for greater awareness of these issues within primary care.
- The provision of reliable information and materials is vital, both for healthcare professionals and patients, to reduce misinformation and fear.
- Physiotherapists with knowledge of Joint Hypermobility Syndrome and Ehlers-Danlos Syndrome Hypermobility Type were cited as sources of support and hope, which helped people to cope with and manage their condition.

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Abstract

Purpose: Little attention has been paid to psychosocial factors in Joint Hypermobility Syndrome and Ehlers-Danlos Syndrome (Hypermobility Type). This study sought to identify the psychosocial impact by examining participants' lived experiences; and identify characteristics of effective coping.

Materials and methods: Adults with Joint Hypermobility Syndrome and Ehlers-Danlos Syndrome (Hypermobility Type) were invited to discuss their own lived experiences and the impact of the condition. All met recognised criteria for clinically significant joint hypermobility, and had a self-confirmed diagnosis. The transcripts were coded and analysed using inductive thematic analysis.

Results: 17 participants (14 women, 3 men) purposively selected to broadly represent different genders, ages and ethnicities. Analysis identified five key themes: healthcare limitations, a lack of awareness of Joint Hypermobility and Ehlers-Danlos Syndrome (Hypermobility Type) among healthcare professionals; a restricted life; social stigma; fear of the unknown; and ways of coping.

Conclusions: The results highlight the significant psychosocial impact on participants' lives. Coping approaches identified included acceptance, building social networks, learning about joint hypermobility and adapting activities. Physiotherapists supported regular exercise. Further research should consider potential interventions to improve information provision, address psychological support and increase awareness of hypermobility among healthcare professionals.

Keywords: Ehlers-Danlos Syndrome, Joint laxity, familial, Joint Instability

, Psychosocial Support Systems, Qualitative Research.

Introduction

Joint Hypermobility Syndrome (JHS) and Ehlers-Danlos Syndrome, Hypermobility Type (EDS-HT) are heritable disorders of connective tissue characterised by generalised joint hypermobility, soft tissue laxity and joint pain [1]. International classification for Ehlers-Danlos Syndromes has recently been revised, with the terms Hypermobile Ehlers-Danlos Syndrome (hEDS), and Hypermobility Spectrum Disorder (HSD), replacing EDS-HT and JHS respectively [2]. However, as this research was conducted prior to the revised nosology, for consistency we will use the combined term JHS/EDS-HT, except where authors have used one term specifically. A previous diagnosis of JHS/EDS-HT was based on medical and family histories in addition to symptoms at clinical presentation, as the genes responsible for JHS/EDS-HT have not been identified [3, 4]. The most common form of EDS, EDS-HT (formerly Type III) has been considered to be the same as JHS; the terms are used interchangeably throughout the literature [3, 4].

Estimates for the prevalence of JHS/EDS-HT are variable within the literature. When examining the prevalence of JHS in patients attending a UK musculoskeletal triage clinic in London, 30% of all participants screened were found to meet the diagnostic criteria for JHS [5]. Literature estimates the prevalence of EDS as approximately 1 in 5000 [6] but due to changes in diagnostic criteria and naming of the syndromes over time the actual frequency of JHS/EDS-HT within the population has yet to be established [7].

The effect of JHS/EDS-HT on people's lives is substantial. Fatigue has been found to be significantly more prevalent in patients with JHS/EDS-HT, compared to the general population [8]. Pain, due to chronic joint pain or injury, can cause severe progressive deterioration of physical functioning and quality of life [7, 9]. In an online survey of 466 adults, JHS/EDS-HT was found to be associated with a number of

comorbid conditions, including anxiety (73%), depression (69%) and fibromyalgia (42%) [10].

A recent review of qualitative literature indicated few studies examining the lived experience and resulting psychosocial impact of JHS/EDS-HT on participants [11]. In Sweden, Berglund and colleagues [12] interviewed eleven interviewees with EDS, recruited from a support group. Results indicated the main theme “living a restricted life” comprised participants’ descriptions of living with pain, living with fear, feeling stigmatised and experiencing of a lack of support when consulting healthcare professionals [12]. In other studies, participants reported being ignored and belittled by doctors, being accused of hypochondria or Munchausen's syndrome, or treated as a spectacle or objectified [13, 14]. Others reported limited social participation due to fatigue, chronic pain or the risk of injury [15]. The impact of JHS/EDS-HT was also felt in the workplace, whether needing to work part-time, making adaptations, or taking early retirement due to symptoms [16].

Management of JHS/EDS-HT is primarily focused on physiotherapy and exercise [17, 18]. However, qualitative studies examining patients’ views of physiotherapy for JHS indicated that referral to physiotherapy services could be difficult and time-consuming [17]. Physiotherapists could struggle to know which exercises to recommend for JHS/EDS-HT, and recommended exercises could sometimes make joint pain worse [17, 19].

In order to gain a greater understanding of the multifaceted impact of JHS/EDS-HT it is necessary to explore a greater number of factors than have previously been studied using biomedical approaches. Despite awareness of high levels of anxiety, depression and low quality of life associated with JHS/EDS-HT [7, 20, 21] within the literature there has been little consideration given to the psychological, social and

cognitive impact of JHS/EDS-HT. In addition, a new toolkit for general practitioners to support management of JHS/EDS-HT in primary care did not consider psychological or social factors [22]. Understanding of these issues is vital in order to design effective interventions to support those with JHS/EDS-HT.

Therefore, the overall aim of this study was to identify the psychosocial impact of JHS/EDS-HT by examining participants' lived experiences, and to identify characteristics of effective coping with JHS/EDS-HT.

Methods

Methodology

Qualitative research was chosen in order to gain a holistic, person-centred perspective from people with JHS/EDS-HT, allowing researchers to generate detailed accounts that give an active representation of that person's reality [23]. In light of the potentially personal and in-depth nature of the research, where participants were anticipated to divulge information about the psychosocial impact of the condition on their lives, potentially including the recall of psychologically challenging experiences such as depression or anxiety, individual semi-structured interviews were chosen over focus groups. Semi-structured interviews permit the examination of social and personal matters in great depth, and can provide researchers with a significant advantage when exploring sensitive topics [24, 25]. Two authors (SB and SP) independently assessed the quality of the final manuscript using the consolidated criteria for reporting qualitative research (COREQ) framework; a 32 item checklist for reporting interviews and focus groups (see Supplementary Table 1) [26].

Ethical approval

Approval for the study was obtained from the University of the West of England Faculty Research Degrees Committee (HAS.16.06.161), and the West London and Gene Therapy Advisory Committee (GTAC) Research Ethics Committee (16/LO/L511).

Hypermobility

Diagnoses in this study were self-confirmed, due to the remote telephone-based nature of the research. The Hakim and Grahame (2003) Five-item self-report questionnaire was used to screen for clinically significant hypermobility, as when a cut-off score of ≥2 is applied it has high sensitivity (80-85%) and specificity (80-90%) to the cut-off score for hypermobility assessed physically (a Beighton score of 4 out of 9) [27].

Participants

Adult participants were recruited through 1) advertisements on the Twitter and Facebook pages of the Hypermobility Syndromes Association (HMSA) and Ehlers-Danlos Support UK (EDS-UK) and 2) recruitment through a local NHS Trust. Eligible participants were aged over 18, with a self-declared diagnosis of JHS, EDS-III or EDS-HT, (due to variations in categorisation of Ehlers-Danlos Syndrome over time), who were able to understand and communicate in English, and gave informed consent. Participants with fibromyalgia were not excluded, as those with JHS are significantly more likely to self-report a diagnosis of fibromyalgia [28]. Respondents with a diagnosis of a different subtype of EDS or any other musculoskeletal disorder were excluded. Participants were purposively sampled using a sampling frame to select from particular representative characteristics, which can be particularly valuable in under-researched populations [29] including age, gender, ethnicity and their five-item

hypermobility questionnaire score [25]. Pseudonyms have been used throughout to ensure confidentiality.

Data collection: Conducting interviews

Prior to study commencement, eligible participants were emailed to arrange the interviews at a time convenient to both parties, with the interview referred to as a 'friendly chat' to reduce anxiety. Telephone interviews were conducted in a private office at the University of the West of England, Bristol between August 2016 and March 2017 by the first author (SB), a female PhD candidate with previous training and experience as a telephone-based emotional support helpline volunteer. Recording was achieved through use of a Dictaphone and in-line recording adapter, with participants reminded that the call was being recorded at the start of each interview. To put each participant at ease, the format and aims of the interview were explained, anonymity assured, and each participant was given the opportunity to ask any questions before the interview questions commenced [30, 31].

Reflexivity

As the first author (SB) has personal experience of EDS-HT and Postural Tachycardia Syndrome (POTS), this was made this clear to participants at the start of the interview. As an 'insider' to the world of Ehlers-Danlos, SB wanted to alter the power imbalance so often seen in traditional research [32, 33]. SB chose to reveal that she had EDS-HT, but emphasised that her experiences were likely to be very different from the participants, and it was their own stories that mattered. Westmarland [34] argued that sharing information about the self and reducing power can work towards creating a more relaxed experience. By acknowledging SB's dual identity both as a researcher and

an individual with EDS, this appeared to instil greater confidence in participants and they seemed very willing to share with SB, a fellow 'in-group' member.

Data collection and analysis

The interviews followed the interview protocol outlined in Supplementary Table 2. These questions were developed using issues highlighted in previous JHS and EDS-HT literature as a guide, in addition to the results of a systematic review and thematic synthesis examining all qualitative JHS and EDS research to date [11]. Due to the exploratory nature of the study, the questions remained broad to allow participants to share their experiences. Questions were pilot-tested by the first author prior to the study, approved by the research team and a patient research partner, and refined as required during data collection to ensure clarity.

The data were analysed by the first author using inductive thematic analysis (TA) as outlined by Braun and Clarke [35] and the analysis was reviewed and discussed with the other authors to ensure consensus. Inductive TA is a very flexible method that allows for a broad focus on meaning across a dataset, and is particularly suited to exploratory study [33]. The transcripts were double-checked for accuracy against the audio recording before being imported into NVivo 10 (QSR International, Melbourne, Australia). This software allowed a better, more efficient overview of the data, and allowed full and equal attention to each data item [35]. Initial data coding took place on an ongoing basis. When no new codes could be identified, data saturation was discussed and agreed with the other authors, and interviews stopped at 17 participants. In this case, saturation was defined as the point where similar answers were offered by participants to the questions, and the when analysis of additional data did not lead the researcher to identify any new codes [36, 37]. Once all the data had been initially coded

and organized, the codes were revised and re-organised into themes, which were derived inductively from the data. Different codes were combined to form themes, and this process revisited and revised, with some irrelevant and minor codes discarded, while others were promoted to overarching themes.

Results

Three hundred and eleven people responded initially, and of these, 145 participants responded to a request to complete the Hospital Anxiety and Depression Scale (HADS) [38]. Five of these did not meet the inclusion criteria, due to secondary medical conditions such as Marfan's Syndrome. 140 participants were screened using a HADS cut-off score of 15. The score allowed participants with clinically significant anxiety or depression to take part, but prevented participants with severe expressions of either condition from being included (scores >15) [38, 39]. This excluded 26 potential participants, or 18.57% of the sample. A purposive sampling strategy was implemented for the 114 remaining prospective participants, where persons were purposively sampled based on criteria relevant to JHS and EDS-HT research. For each category (age, gender, ethnicity, 5PQ score and levels of anxiety and depression as measured by the HADS) participants in the list were randomly selected using a custom-range true random number generator.

A total of 17 people (14 women, 3 men) took part in the study. All had a diagnosis of JHS/EDS-HT or EDS-III. Five participants (4 women, 1 man, 29.4% of the final sample) also had a diagnosis of Fibromyalgia, which is comparable to rates of Fibromyalgia in other JHS populations (27.3%, [28]; 30%, [40]). Twelve participants identified as 'White', four identified as 'mixed White and Asian', and one participant as 'mixed: White and Indian'. Five participants were from the South-East of England, three from the South-West, three from the Midlands, one from the North-West, three

from the North-East, and two from Scotland. Interviews lasted between 43 and 99 minutes (with a mean duration of 74 minutes). In the quotes below, pauses in speech are indicated by "(.)", while ellipses "…" indicate omitted text.

Five overarching themes were identified through analysis of the interview data:

(1) Restrictions imposed by JHS/EDS-HT
(2) Healthcare limitations
(3) Social stigma
(4) Fear of the unknown
(5) Ways of coping

Each major overarching theme featured data from all 17 participants. These are presented below, with illustrative quotes as appropriate. Data illustrating theme and subtheme frequency counts to indicate the representativeness of the findings across the data is listed in Supplementary Table 3. To ensure participant confidentiality, pseudonyms have been used throughout.

Theme 1: Restrictions imposed by JHS/EDS-HT

Many participants described limitations in their daily activities due to factors such as frequent dislocations, restricted mobility and symptoms of fatigue and pain. The patellae, shoulders, jaw and hips were cited as the most often dislocated, but potentially any joint was at risk. Problems with swallowing and digestion limited participants' ability to maintain a 'normal' social life. The majority had problems completing housework, cooking, shopping or dressing.

Some participants also reported difficulties with sex and intimate relationships. These problems seemed to stem from issues around fatigue and pain, as Nigel explains: "*I'm always so tired, or I'm aching somewhere, you can't really get in the mood….*" Jake found it "*very difficult … to talk about things like erectile dysfunction"*, later reflecting that there was little his doctor could do to help. For women, gynaecological complications such as prolapse could have a similarly distressing impact, as Jackie said: *"You don't want anyone to come anywhere near you*". These difficulties led to worry regarding relationships with their partners. Nigel was fearful that his wife might leave him: "*Is she going to be understanding …*[for] *The rest of our time together?… It's a big thing, not having…the sexual side of a relationship*".

Relying on others

To complete the tasks that they couldn't manage, participants sometimes had to rely on others for support. However, this left them feeling guilt, like *"a burden"* [Roger], with fears that they were *"restricting"* [Roger] or holding back [Frances] their family members. Over time, worries about their responsibilities could lead to exacerbations of anxiety and depression, as Roger describes: "*Sometimes I feel a burden, or a nuisance… that's why I end up…having the anxiety problems … feeling sorry for other people, having to put up with me".*

Work life and education

The majority of participants had started their working lives without a diagnosis of JHS/EDS-HT. Many had initially worked in roles that were not suited to their condition and were forced to re-think their career options. In an effort to manage fatigue, participants worked reduced hours, or from home. Working was cited as very important to participants; they felt that work gave them a role in society and a purpose in life,

which in turn benefited their emotional wellbeing: "*Even if I don't get (.) any money or anything, at least I'm doing something useful*" [Rachel]. Anna emphasised the need to stay positive, despite the potential restrictions to her work life: "*for my wellbeing I feel that I need to be doing something…So I try to keep working…to try and keep myself positive".*

Theme 2: Healthcare limitations

Lack of awareness of JHS/EDS-HT

Every participant interviewed reported an overall lack of awareness of hypermobility syndromes such as JHS/EDS-HT among healthcare professionals, including consultants, General Practitioners (GPs), nurses and physiotherapists. This naturally resulted in patients waiting many years to be diagnosed. Others were labelled as "*hypochondriac"*, several as "*making it up"*, some as the symptoms being "*all in your head"*, or accused of having a mental health problem. However, after many instances of being mislabelled as hypochondriacs, several participants described their diagnosis as a positive, validating experience, as years of symptoms were finally given a recognized cause: *"…it all makes sense now… you realise you're not going mad!"* [Claire].

Just bendy joints

However, even with a verified diagnosis, some felt that their symptoms were still dismissed by healthcare professionals as, *"just bendy"* [Mandy], rather than seeing the wider psychosocial impact of the condition on participants' lives. One participant, Mandy, reflected on whether her consultant's attitude was due to a lack of understanding about JHS/EDS-HT: *"Even if they've heard of EDS, it's misunderstood…'Yeah, you got bendy joints' … And it's like, 'Yeah, but there's a bit*

more to it than that!'".

Almost all participants gave examples of healthcare professionals' negative reactions and attitudes, such as becoming angry or being dismissive of their symptoms. Others spoke of feeling objectified during medical examinations, with their hypermobility treated as a spectacle. Frances gives an example:

> *...Then* [Doctor] *was like, 'Wow! You're such a freakshow!', and I'm like (.) 'Wow. Not a nice thing to say, thanks!'* …[He said] '*Ooh, let's get all the medical students here and show them!' And I'm like, 'No!'*

Limitations of current treatment

A significant number of participants described instances where local anaesthetics had been ineffective due to their hypermobility syndrome. This can result in patients undergoing surgical and dental procedures fully aware of pain: *"I could feel what they were doing. And I- I spoke up, and (.)* [the doctor] *basically said, 'No, you can't.' and carried on… but I was in a lot of pain?"* [Mandy].

Nigel and Bryn had both made attempts to warn their surgeons during subsequent procedures, but to little success:

> *"and the surgeon said, 'What's Ehlers-Danlos?'"* [Nigel]

> *"They* [had] *no idea"* [Bryn].

A combination of dismissive attitudes from healthcare professionals, and a lack of treatment options had made Rachel wary: *"I just want to keep away from doctors if I can… unless its desperate I won't bother.".*

Theme 3: Social Stigma

Difficulty keeping up

Difficulty in keeping up with friends, family and colleagues led to feelings of frustration and anger in participants, because their joints could not always endure what they wanted to do, as Nigel describes: *"Why can't I do what everybody else does? Why am I the only one who can't (.) Go for this walk?' You know? 'Do this thing that everyone else is doing?".*

Participants described a number of internalised negative feelings about their own bodies. Several described themselves as *"weird"* or *"useless"*, experiencing shame and guilt. Rhiannon expressed a longing to be "*normal*": *"I guess made me (long pause, 3 seconds) a little bit? Frustrated with my body? That it can't be a bit more normal.".*

Judgements of others

Due to the invisible nature of JHS/EDS participants often faced judgements from friends, family and strangers, as Jackie states: *"I've also gotten abuse off of disabled people? …'why are you parking in a blue badge space?'".*

Many described feelings of embarrassment, guilt and shame when confronted by others. Participants mentioned '*you look well'* [Tabitha], '*I'm a younger person'* [Claire], '*I'm the one who looks less disabled'* [Dana] or '*you look fine'* [Nigel] as potential reasons why they had been stopped. Mandy questioned whether this judgement was due to those with JHS/EDS-HT not fitting the common perception of disability as a visible difference: *"But, other people's judgement…if you don't fit that (.) Notion of what disabled is in their head, then they get really confused.".*

Hiding symptoms from others

Fears of being judged led many to hide their symptoms, in an effort to "*pretend it's okay*" [Nigel]. Participants expressed hesitancy in being perceived as '*faking things'* [Jackie] or '*as if you're complaining the whole time* [Tabitha]. Lauren mentioned a need to *"Try and appear normal all the time"*. Many believed that other people might think negatively of them if they knew about their JHS/EDS-HT. Divulging their invisible difference to others took a great deal of confidence, yet others described feeling embarrassed when asking for accommodations such as a seat on a busy train, *"People are judging me.' and I just kind of want to (.) get on quietly and I'll skulk away"* [Mandy].

Men in the present study felt that the need to ask for seats had 'chipped away' at their masculinity. Some wondered whether their JHS/EDS-HT would be more readily accepted by others if it were more visible, as Jackie recounted: *"I actually wish I was in a wheelchair! Do you know what I mean? I just feel like (.) People can understand that?"*.

Numerous participants found the invisible nature of their condition an asset, preferring not to '*let* [JHS/EDS-HT] *define me'* [Wendy]. Similarly, other participants felt that they could choose when to bring up their disability, and how. Choosing what and when to tell others about their condition afforded participants a sense of control. However, the effort of hiding differences from others can be psychologically demanding over time.

Theme 4: Fear of the unknown

Fears of future decline

Not knowing when the next injury was going to occur, how their illness trajectory was

going to affect them over time, or the potential impact of JHS/EDS on their future plans made participants especially fearful of future declines in their physical ability. Others described feelings of panic that they still had not achieved everything that they wanted to do:

> *It's genetic…we can't fix this, what does this mean for the future?... You know? 'I'm not going to be able to get married, or have kids', all the things that I thought I would.* [Mandy]

For participants with a family history of JHS/EDS-HT, their parents' struggles were a vision of their own future, as Frances described: *"All the things I can do now, [Mum] could do when she was my age…But now, she needs a double knee replacement because her knees are just so worn out?".*

Fears of future decline linked to Roger's worries about depending on other family members, and feelings of guilt, shame and being a burden on his loved ones: *"I still am- frightened of what the future holds... one day I might not be able to do anything. And it's that fear of being a burden again?".*

Many participants took the initiative to find out more about their condition, and had joined support groups and a variety of social media pages. However, seeing others with JHS/EDS-HT, in person and online, who were more severely affected than themselves sometimes led to a cycling progression of fears concerning prospective future declines in ability. These fears led to fearful responses to new or unusual symptoms, as participants became anxious as to whether the new symptom signalled their own health decline:

> *Some of the stuff on the internet…- it fills your head with fear in some respects… and I'm like, 'Ah! I don't want to end up like that!'. It's that fear that, 'Ah, that could happen!'…over-thinking things and then I start panicking, it's like, then it's a vicious circle*. [Roger]

While others felt that social media could lead to competition: "*Even in the EDS community… People almost sort of, trying to outdo each other, with, 'Oh, look how ill I am, look how it impacts on my life.'*" [Tabitha].

Lack of reliable information

Many spoke of a lack of reliable information about their condition. All accepted the need to gain information about JHS/EDS-HT from reputable sources such as patient support groups, rather than "*just picking up things on forums*" [Roger]. However, participants recognised that not everyone with JHS/EDS-HT would have access to trustworthy information: *"And I think there's a lot of… misinformation out there as well I think …There doesn't seem to be a huge amount of research into it, or information?"* [Rhiannon].

Pregnancy and heritability

Participants' fears for the future also applied to decisions about having children. Due to the hereditary nature of JHS/EDS-HT, participants considering future children were worried about whether their children would inherit JHS/EDS-HT from them: *"But yeah. It's always that fear (.) Of passing it on, and seeing someone else have to go through, what I've gone through?"* [Roger].

Many of the participants in the present study had been diagnosed with JHS/EDS-HT much later in life. Some who had found out about their condition years after having families had faced many issues:

> *Because [EDS-HT] wasn't picked up I've had (.) premature babies and (.) extensive problems during pregnancy, life-threatening miscarriage and things like that… if I'd known the complications, and the risks, of having a baby, then I probably wouldn't have gone on to have four.* [Anna]

Others struggled with the guilt of having unknowingly passed JHS/EDS-HT on to their children: *"If I'd have known, I wouldn't have inflicted this on anyone."* [Lauren]. Mandy described the guilt felt by her mother: *"When I first got diagnosed, I think my mum had felt quite a bit of guilt? Um, partly the (.) Hereditary thing, thinking she's passed it on."*

Perceived lack of psychological support

Some felt that psychological support to better cope with the stress and enduring psychosocial impact of JHS/EDS-HT on their lives was lacking:

> *"What I would say is that nobody … considers the psychological impact… I got diagnosed, and then was basically just told- …'Well, this is just going to get worse.' And then you get sent away! And no-one seems to think they've just given you a life-changing diagnosis.* [Jackie]

> *…how it affects you emotionally… I think that can get forgotten by doctors, sometimes…or at least it's not looked at as a whole package, necessarily."*[Wendy]

Neither Jackie nor Wendy felt that the psychological or emotional impact of JHS/EDS-HT was taken into account. Others wished for support when stressed, depressed or anxious; to address their fears about the future and worries about potential declines in their condition.

Theme 5: Ways of coping

Psychosocial and cognitive appraisals: Acceptance

Many cited acceptance of the life-long nature of their condition as having a beneficial impact, and acknowledged receiving a diagnosis as part of that. By recognising the need to pace activities and "*stay in sometimes because I just can't (.) Do everything.*"

[Georgina], participants were better able to manage the psychosocial impact of JHS/EDS-HT on their lives. As Frances stated: *"I think now, I'm like, 'Do you know what? I'm weird, and I'm bendy, and I'm always going to be that way, nothing is going to change how I am now.'".* Acceptance of a chronic condition, including its implications and limitations, seemed to give participants a sense of control. Numerous participant cited the need to be optimistic, and staying '*positive'* in the face of repeated flare-ups and setbacks was also emphasised: *"Okay, that's how I am at this point in time, but tomorrow is another day…do something that makes you smile."* [Lauren]. By downwardly comparing themselves to others who had more severe JHS/EDS-HT or other life-limiting conditions, many participants felt that life "*could always be worse"* [Roger] and chose to see the positives in their own situation: "*'I'm still fine, I'm still alive, there are worse things that I could have', you know?"* [Frances]. Although JHS/EDS-HT had limited Frances' ability to '*keep up'* with her friends socially, she found that she could "*still do stuff that I enjoy and have fun".* Many spoke of the need to harness their determination, to "*find a way"* to complete the goals they had set their mind to. As Jackie explained: "*rather than just accepting 'no'. 'Is there a different way I can do this?'".*

Social support

As friendship groups shifted, many participants made friends with, and gained social support from people with JHS/EDS-HT or other chronic health conditions. Participants perceived these 'similar' friends as more understanding and empathetic to what they were going through. As Georgina put it: *"…the friends I do have now are the ones that are similar to me, not necessarily hypermobile but they've got mild disabilities. So they, at least, can relate."*. The combination of social support and finding out more about JHS/EDS-HT gave participants the confidence and assertiveness to explain and educate

others about their condition when challenged:

> So yeah, [when people don't understand] doesn't particularly frustrate me. I see it as more of a chance to educate people about EDS and other invisible illnesses. I'm really happy to talk about it. As I say, you start me off on it and I never shut up! [Wendy]

Physical and Behavioural :Hobbies and projects

Participants cited various hobbies and projects as a way to keep themselves '*sane'* [Nigel]. Nigel had always really enjoyed DIY and got great enjoyment from hands-on building projects, but the impact of such activities left him *'completely ruined'* for weeks after. Later, he found a good substitute to give him the same level of happiness and satisfaction, which he felt occupied his mind from '*dwelling on things'*:

> *It's Lego… because I can do that sitting down, I haven't got to, you know, expend too much energy by doing it…it's helped my- my self-esteem, and my mental side of it, more than anything*. [Nigel]

Positive interactions with healthcare professionals

Although participants reported that some recommended exercises made their pain worse, others had reported very positive experiences of physiotherapy. These were physiotherapists who had specialist JHS/EDS-HT knowledge, who could provide emotional support and encourage regular exercise:

> *She's shown me techniques … how to put* [subluxations] *straight back before it comes out and things like that. So I'm not as, frightened in some respects? There's a bit more light at the end of the tunnel.* [Roger]

While they may not always recognise the condition, participants were pleased when clinicians were willing to learn: As Jake put it, "*it's more… how they respond to NOT knowing, than what they know.*" While it was acknowledged that GPs could not be

expected to know everything about rare genetic disorders, participants felt that greater training and awareness of JHS/EDS-HT among healthcare professionals could improve outcomes for patients.

Often, it was not that a treatment had worked, but that healthcare professionals had taken the time to listen compassionately and empathetically to patients "*rather than just (.) Pacifying you, and sending you away*." [Mandy]. By acknowledging the holistic, multidisciplinary impact of JHS/EDS-HT and providing useful guidance, healthcare professionals could prevent participants from feeling that their condition was not recognised.

Discussion

This study provides a novel understanding of how psychosocial factors, such as the support of family, social networks and attitudes of healthcare professionals are perceived to play significant roles in shaping men's and women's experiences of JHS/EDS-HT.

Invalidation can be a common problem with rheumatic diseases [41], and involves a lack of understanding or negative social responses from others, such as disbelief, rejection, stigmatization and suspicion that the problem may be psychological in origin [41]. In line with other examples within the literature, participants in this study experienced very long waits for diagnosis, allegations of hypochondria or malingering, and a lack of understanding and knowledge of the condition from healthcare professionals [12, 13, 15, 42, 43].

Pain and fatigue, the main symptoms in JHS/EDS-HT, are mostly non-observable by others; therefore participants' symptoms and the associated burden of the condition were often cited as being poorly understood. Participants in the present study hid their symptoms in an attempt to appear 'normal' and avoid confrontation. Shame,

guilt and stigma can have negative psychosocial consequences, lowering self-esteem and in some cases leading to depression [44]. Interventions to address issues around self-esteem in chronic pain populations have targeted social functioning, including issues such as as anger management, dealing with depression, uncertainty and self-esteem [45, 46]. Therefore, future psychosocial interventions and support to address feelings of stigma and improve self-esteem would be valuable.

Others recounted significant pain from reduced effectiveness of local anaesthetics, thought to be due to the underlying collagen defect in JHS/EDS-HT [47]. These negative experiences could lead to a fear of treatment, which may prevent those with JHS/EDS-HT from seeking appropriate medical care [12]. When finally diagnosed, participants reported feeling great relief at understanding their condition, in line with JHS/EDS-HT research [14, 43] and other difficult-to-diagnose conditions [48]. Some progress is already being made towards improving awareness of JHS/EDS-HT amongst general practitioners through a newly published EDS GP Toolkit, in partnership with the Royal College of General Practitioners (RCGP) and a patient support group, Ehlers-Danlos Support UK (EDS-UK) [22]. However, there is still an overall lack of training and awareness of JHS/EDS-HT among GPs and healthcare professionals, and the guidance does not cater for the psychosocial impact of the condition.

In addition to difficulties with local anaesthetics, recent important research by Rombaut et al [49] and Scheper and colleagues [50] also indicated significant hypersensitivity to pain in patients with JHS/EDS-HT, compared to the general population. Generalised hyperalgesia, defined as an abnormally increased sensitivity to pain, has been tested using pressure-pain thresholds, (PPT's) at a range of body areas in patients with EDS-HT compared to control participants [49, 50]. Results revealed that those with EDS-HT to have significantly lower thresholds compared to the control

group, even in non-symptomatic areas of the body, indicating generalised hyperalgeisia [50]. These results suggest the possibility of central nervous system sensitisation, hypothesised as being due to the collagen defect affecting the axonal function of peripheral nerves in JHS/EDS-HT, which could leave this population more prone to chronic pain and fatigue [8, 49]. Therefore, the tailoring of multidisciplinary pain management interventions, including cognitive behavioural therapy and physiotherapy would be an important consideration, in order to reduce overall disability and improve patient quality of life [49].

Participants with JHS/EDS-HT experienced numerous restrictions to their lives as a result of a range of symptoms, including pain, fatigue and the unpredictability of their condition. Due to the multisystemic nature of JHS/EDS-HT, participants can experience a number of additional health problems such as functional gastrointestinal issues, urinary problems and autonomic dysfunction [51]. These additional multifactorial symptoms, in combination with environmental factors such as social stigma or isolation may contribute to psychological distress and disability in this population [52].

Issues around the effects of pelvic organ prolapse in JHS/EDS-HT echo a recent questionnaire survey which found more severe sexual dysfunction and a greater negative impact on quality of life in women with JHS compared to the general population [53]. Difficulties with sexual relationships due to vaginal or bladder prolapse in women, or erectile dysfunction in men are an under-researched finding; participants feared a negative impact on their relationship, and were unsure of what could be done medically to solve their problems. Erectile dysfunction can also be a symptom of autonomic dysfunction, a common co-occurring diagnosis in JHS/EDS-HT. Autonomic dysfunction has also been linked to sexual dysfunction in Parkinson's disease [54].

However, there has been little assessment of this issue within the JHS/EDS-HT literature. In JHS/EDS-HT research populations women typically outnumber men, and in a recent study of autonomic dysfunction symptoms, the majority of men with EDS-HT did not want to complete the erectile dysfunction symptom profile, so its impact was not recorded [55]. These results indicate the need for greater support and recognition of potential intimacy issues in JHS/EDS-HT, particularly for health professionals in primary care, as problems with sexual functioning may markedly influence the patient's quality of life.

A scarcity of information and dependable psychological support for JHS/EDS-HT led some to be very fearful of future declines in their ability. In addition the potentially negative influences of finding information on the internet were also highlighted by participants, and are a new consideration in this area. Several participants described these fears as leading to catastrophizing and feelings of panic when faced with new symptoms, in case this signified their own decline [56]. In addition, when people make social comparisons to others in the same chronic illness group there is a risk that other support group members' decline could be interpreted as a representation of their own future, resulting in negative feelings [57, 58, 59, 60]. Due to the hereditary nature of the condition, this is particularly relevant to children whose parents also have JHS/EDS-HT. Psychosocial factors such as fear-avoidance behaviours, activity limitation, catastrophizing and fear of movement may substantially contribute to increased levels of disability in this patient group, as has been seen in other chronic pain populations [61, 62]. In this case, maladaptive beliefs relating to JHS/EDS-HT (such as the belief of a sudden decline) appeared to lead to heightened emotional reactivity, leading to catastrophizing, anxiety and feelings of panic [62]. In order to counteract this effect, it may be beneficial for support group members to be

provided with opportunities for positive social comparison, as interaction with other patients has been shown to help reduce fear relating to symptoms [63]. Modelled behaviour involves providing example behaviours for people to aspire to or imitate [64]. With links to Bandura's Self-Efficacy Theory, self-efficacy relates to an individual's expectations and beliefs about their ability to perform specific actions effectively [65]. Video modelling, or the demonstration of desired behaviours through visual media, has been used successfully in patient education and to facilitate learning of new skills [65], including prostate [66], breast [67] and colorectal [68] cancer screening, and adherence to self-care behaviours for participants with heart failure [69]. By using models to promote exemplary behaviours, video modelling can serve as a useful way to promote self-efficacy in others [70]. For instance, examples of self-help ideas and positive coping strategies could be promoted by those with JHS/EDS-HT using video clips, for people experiencing a flare in symptoms.

These results also demonstrated the physical and psychosocial impact of JHS/EDS-HT in terms of adjustment and adaptation to the condition, extending knowledge regarding coping strategies employed by those with JHS/EDS-HT. Initially, diagnosis was met with relief, but later reactions indicated that participants had to face changes in how they saw themselves, and how others perceived them, reflecting a period changes to their identity. These results are similar to other research indicating that chronic illness and disability can bring about great changes in a person's identity [71]. Many cited acceptance of JHS/EDS-HT as positively influencing their ability to cope. Acceptance in this case can be defined as refocusing attention on aspects of the condition that they can control, rather than struggling to control pain [72]. These findings are similar to those of other studies, which found that acceptance promoted adjustment to chronic disease [73, 74, 75]. In addition, rheumatic diseases research has

indicated that acceptance of a pain condition, in addition to high self-efficacy, can have a buffering effect against invalidation by others, such as healthcare professionals, friends or family [76]. Participants who identified a need for greater psychological support indicated that this should be available as soon as possible following diagnosis in order to better support this process and transition.

Building social networks and social support were also cited as useful coping strategies by participants, including joining support groups and making friends with others who had JHS/EDS-HT. Online and face-to-face support communities play an important role for those with chronic diseases in conveying information, gaining emotional support and comparing experiences [77, 78]. Social comparison theory may be useful in understanding how these comparisons influence quality of life. Downward social comparison, the comparison of the self to someone doing less well, has been associated with improved quality of life [79], and can be used to enhance positive affect [80]. By gaining knowledge about their condition, participants were able to foster a sense of control, and lessen the feelings of '*unknown'* and fear that can be present in unpredictable conditions [75, 81].

In line with prior research [12, 15, 16] participants described how becoming dependent on others for help with activities could result in feelings of guilt and shame. This highlights a need for greater support for participants with JHS/EDS-HT in order to maximise independence in everyday activities, particularly in relation to personal care, washing and dressing. This is a key indication that rehabilitation and support for people with JHS/EDS-HT needs a greater focus on maximising independence in activities of daily living.

Many participants actively adapted hobbies and sports to better achieve their ambitions. Healthcare professionals such as physiotherapists with knowledge of

JHS/EDS-HT were cited as helping them to achieve their goals. By giving patients an indication that things could improve, physiotherapists gave patients hope. Encouraging patients to adjust to daily life may lead to a sense of control and stimulate active coping strategies. A recent qualitative study of physiotherapy for JHS recommended a holistic approach to management, rather than treating single joints in isolation [17]. Specialist physiotherapists with knowledge of JHS were cited as being very helpful. However, specialist physiotherapy is limited in the UK, and there is little consensus regarding optimal exercise [82]. Further exploration of patient and healthcare professionals' ideas about optimising supportive interventions for JHS/EDS-HT would be valuable.

Strengths and Limitations

This is the largest study of participants' lived experiences of JHS/EDS-HT, compared to previous semi-structured interview studies (n=10, [15]; n=11 [12, 16]). In addition, this study had a proportionally higher representation of participants of mixed ethnicity. While hypermobility is known to affect Black and Asian populations to a greater extent [5], very few ethnically diverse participants have been involved in JHS and EDS research compared to participants of White ethnicity. While this study purposively sampled to gain a broader representation, further work is still required to assess whether any new themes would be identified with greater representation of black and minority ethnic populations, whose views in JHS/EDS research have yet to be explored in great depth. In addition, while men were involved in this study, their experiences of JHS/EDS-HT compared to women have yet to be fully studied.

In addition, while the position of the first author (SB) as both a patient and a researcher could be considered a strength, the authors were also aware of the possibility of bias as a result of this dual position. While bias cannot be eliminated entirely, it can be mitigated [83]. In order to counteract this, the first author (SB) kept a reflective practice

research diary exploring her feelings and reflections after each interview. A random selection of recordings was checked against transcripts by the research team (NW, TM, SP), thereby allowing those not directly involved in data collection to audit the results, reducing potential bias and ensuring accuracy [83]. In addition, peer debriefing was used to safeguard externality, where the research team (NW, TM, SP) reviewed the findings and themes identified in the results. This allowed the first author the chance to think more critically about the research, and to discuss and explore judgements made about the data.

Implications for clinical practice

These findings have built upon previous findings in this area, including sexual dysfunction [84], and requirements for improved awareness among healthcare professionals [13, 14, 16, 43]. New topics have also been established, such as a need for greater independence in activities, fear regarding symptoms and decline, and the risk associated with social comparisons to others with JHS/EDS-HT. The importance of social support, patient groups and communities was highlighted, as was the need for future interventions to improve feelings of stigma and boost self-esteem. It would be helpful to map what is now known about the psychosocial impact of JHS/EDS-HT to behaviour change theories, such as the Theoretical Domains Framework or COM-B [64, 85]. This could help to develop focused behaviour change interventions based around factors identified by patients as promoting or hindering their ability to cope with JHS/EDS-HT.

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Declaration of Interest Statement

The authors declare no conflict of interest in respect to the authorship of this article.

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Supplementary Table 1: Consolidated criteria for Reporting Qualitative Research (COREQ) checklist.

Tong, A., Sainsbury, P., & Craig, J. (2007). Consolidated criteria for reporting qualitative research (COREQ): a 32-item checklist for interviews and focus groups. *International Journal for Quality in Health Care, (19)*6, 349-357.

Topic	Item No.	Guide questions/description	Reported on page number/section
Domain 1: Research team and reflexivity			
Personal characteristics			
Interviewer/facilitator	1	Which author conducted the interview or focus group?	Data collection: Conducting interviews
Credentials	2	What were the researcher's credentials? E.g. PhD, MD?	Data collection: Conducting interviews
Occupation	3	What was their occupation at the time of the study?	Data collection: Conducting interviews
Gender	4	Was the researcher male or female?	Data collection: Conducting interviews
Experience and training	5	What experience or training did the researcher have?	Data collection:

			Conducting interviews
Relationship with participants			
Relationship established	6	Was a relationship established prior to study commencement?	Reflexivity & Data collection and analysis
Participant knowledge of the interviewer	7	What did the participants know about the researcher? E.g. personal goals, reasons for doing the research?	Reflexivity
Interviewer characteristics	8	What characteristics were reported about the interviewer/facilitator? E.g. bias, assumptions, reasons and interests in the research topic	Reflexivity
Doman 2: Study design			
Theoretical framework			
Methodological orientation and theory	9	What methodological orientation was stated to underpin the study? E.g. grounded theory, discourse analysis, ethnography, phenomenology, content analysis.	Inductive Thematic Analysis
Participant selection			
Sampling	10	How are participants selected? E.g. purposive, convenience, consecutive, snowball sampling?	Purposive sampling: Results section
Method of approach	11	How were participants approached? E.g. face-to-face, telephone, mail, email	Data collection: Conducting interviews
Sample size	12	How many participants were in the study?	17 participants: Results section
Non-participation	13	How many refused to participate or dropped out? Reasons?	N/A

Setting			
Setting of data collection	14	Where was the data collected? E.g. home, clinic, workplace	Results section
Presence of non-participants	15	Was anyone else present besides the participants and researchers?	N/A
Description of sample	16	What are the important characteristics of the sample?	Results section
Data collection			
Interview guide	17	Were questions, prompts, guides provided by the authors? Was it pilot tested?	Data collection and analysis
Repeat interviews	18	Were there repeat interviews? If so, how many	N/A
Audio/visual recording	19	Did the research use audio or visual recording to collect the data?	Data collection and analysis
Field notes	20	Were field notes made during and/or after the interview or focus group?	N/A
Duration	21	What was the duration of the interviews or focus group?	Results section
Data saturation	22	Was data saturation discussed?	Data collection and analysis
Transcripts returned	23	Were transcripts returned to participants for comment and/or correction?	N/A
Doman 3: Analysis and findings			
Data analysis			
Number of data coders	24	How many data coders coded the data?	Data collection and analysis
Description of the coding tree	25	Did the authors describe a description of the coding tree?	No
Derivation of themes	26	Were themes identified in advance or derived from the data?	Data collection and analysis

Software	27	What software, if applicable, was used to manage the data?	Data collection and analysis
Participant checking	28	Did participants provide feedback on the findings?	No
Reporting			
Quotations presented	29	Were participant quotations presented to illustrate the themes/findings? Was each quotation identified? E.g. participant number	Yes, Results section. Yes, all identified by participant pseudonym.
Data and findings consistent	30	Was there consistency between the data presented and the findings?	Yes, we think so.
Clarity of major themes	31	Were major themes clearly presented in the findings?	Yes, we think so.
Clarity of minor themes	32	Is there a description of diverse cases or discussion of minor themes?	Yes, we think so.

Supplementary Table 2: Interview topic guide used in individual interviews	
Concept	Question
Diagnosis Journey	• Can you clarify for me whether your diagnosis is joint hypermobility syndrome (JHS) or Ehlers-Danlos Syndrome (EDS-HT) (or both)? • How did you come to be diagnosed? How long was it before you first started having symptoms and receiving your diagnosis? • What age were you at diagnosis?
The impact of JHS/EDS-HT on participation	• What are your symptoms? • Do you have physical symptoms e.g. subluxations or dislocations, or fatigue? • How do you manage your day-to-day activities? • Do any activities make your symptoms worse? What happened? • What impact does JHS/EDS-HT have on your education and/or your work life? Has having JHS/EDS-HT had any impact on your relations with other people, such as friends or family? • What impact has JHS/EDS-HT had on your social and leisure activities?
Impact on the individual, or on the perception of the self	• Has having JHS/EDS-HT had an emotional impact? • How do you feel about having JHS/EDS-HT? • Has having JHS/EDS-HT changed how you think or feel about yourself? • Have they been any positive impacts of having JHS/EDS-HT? (e.g. relief at diagnosis?)
Identifying coping	• Is there anything that you have tried that has had a beneficial impact on your condition? e.g. Physical (such as exercises, physiotherapy, Pilates, yoga, gymnastics etc.), or emotional (e.g. relaxation, mindfulness, cognitive behavioural therapy, pain management programmes)? • Can you say what it is about these methods that helped?

	• If you had to give advice to another person about how to cope well with JHS/EDS-HT, what would that be? • Have you taken steps to educate yourself about JHS/EDS-HT and find out more about your condition? Why/why not?
Individual's perceptions of others	• Some research has suggested that JHS/EDS-HT are poorly understood conditions, whereas others have more positive experiences. What are your views on this? • Have you had any difficulties, or benefits, from the 'invisible' nature of JHS/EDS-HT? What have these difficulties or benefits been? • Who were these with (e.g. family, friends, co-workers, healthcare professionals, strangers)?
Experiences of healthcare	• What have been your experiences of healthcare professionals, e.g. physiotherapists, occupational therapists, nurses, GP's, consultants, psychologists? • What kinds of treatment have you been offered? • Did your treatment change following your diagnosis? S • ome research has suggested that healthcare professionals can struggle to know how to treat patients with JHS/EDS-HT, whereas others seem to be more confident, I wonder which is closer to your own experiences?
Closing	• Is there anything else about your experiences, thoughts or feelings of living with JHS/EDS-HT that I haven't mentioned? • Any questions that you thought I would ask, or wished I'd asked, but didn't?

Supplementary Table 3: Frequency counts for each theme and subtheme

Each major overarching theme featured data from all 17 participants.

The frequency counts for coding in relation to each participant are presented below. All major themes featured data from all 17 participants, as did the subthemes 'lack of awareness of JHS/EDS-HT', 'difficulty keeping up', judgements of others'.

Supplementary Table 2: A table to illustrate the representativeness of the findings across the data.

Theme and subtheme	**Number of transcripts coded to the theme or subtheme (n=17)**
Theme 1: Restrictions imposed by JHS/EDS-HT	17
• *Relying on others*	9
• *Work life and education*	11
Theme 2: Healthcare limitations	17
• *Lack of awareness of JHS/EDS-HT*	17
• *Just bendy joints*	12
• *Limitations of current treatment*	12
Theme 3: Social Stigma	17
• *Difficulty keeping up*	17
• *Judgements of others*	17

• *Hiding symptoms from others*	14
Theme 4: Fear of the unknown	17
• *Fears of future decline*	13
• *Lack of reliable information*	7
• *Pregnancy and heritability*	9
• *Perceived lack of psychological support*	3
Theme 5: Ways of coping	17
Coping: Psychosocial and cognitive appraisals	17
• *Acceptance*	12
• *Social support*	15
Coping: Physical and Behavioural	17
• *Hobbies and projects*	14
• *Positive interactions with healthcare professionals*	15