Understanding hypermobile Ehlers-Danlos syndrome and Hypermobility Spectrum Disorders in the context of childbearing: An international qualitative study

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Abstract

Objective

The Ehlers-Danlos syndromes (EDS) and Hypermobility Spectrum Disorders (HSD) have profound and life-threatening consequences in childbearing as they affect connective tissues throughout the body. Hypermobile EDS (hEDS) and HSD are estimated here for the first time to affect 6 million (4.6%) pregnancies globally per year. The aim of this study was to arrive at a deeper biopsychosocial understanding of childbearing in the context of hEDS/HSD.

Methods

English speaking women aged over 18 years who had previously given birth and had a confirmed medical diagnosis of hEDS/HSD or equivalent diagnosis under a preceding nosology were included in this study (n=40). Narrative interviews were used to collect qualitative data from this international sample of participants. Thematic narrative analysis was used to understand how participants made sense of their experiences.

Findings

Participants were aged between 25 and 55. Births (n= 52) between 1981 and 2018 were captured across United Kingdom=29 (73%), United States of America=10 (25%) and Canada=1 (2%). The majority of participants interviewed recounted a worsening of symptoms during pregnancy and postnatal complications. Anaesthesia was often reportedly ineffective, and for many, long latent phases of labour quickly developed into rapidly progressing active labours and births. Maternity staff were observed to be panicked by these unexpected outcomes and were deemed to lack the knowledge and understanding of how to care for women in this context. Poor maternity care resulted in women disengaging from services, trauma, stress, anxiety and an avoidance of future childbearing.

Key conclusions and implications for practice

Cases of hEDS/HSD should no longer be considered rare in maternity services. Maternity staff must be adequately prepared for this new reality. As a first step, www.hEDSTogether.com has been developed to provide a repository of evidence in relation to this topic, along with a freely downloadable toolkit for use in practice. It is important to listen, acknowledge and respond to women with hEDS/HSD appropriately throughout their childbearing journey. Dismissal can lead to trauma and needless morbidity.

Keywords: Pregnancy; Birth; Postnatal; Ehlers-Danlos Syndrome; Joint Hypermobility; Hypermobility Spectrum Disorders

INTRODUCTION

Both the World Health Organisation's recommendations for a positive childbirth experience (World Health Organisation, 2018a) and the United Nation's Global Strategy for Women's, Children's and Adolescents' Health (World Health Organisation, 2018b) aim to achieve safer childbearing, and meet the psychological and emotional needs of all women. New declarations also set out the importance of person-centred care for people with long term conditions (NHS England, 2019). Some diagnosed with Ehlers-Danlos syndromes (EDS) perceive a lack of clinical knowledge and awareness among health care professionals about their condition (Terry et al, 2015). Childbearing for women with EDS is associated with a number of complications such as preterm and precipitate birth, uterine atony, pelvic organ prolapse, haemorrhage, wound dehiscence, poor anaesthetic coverage and tissue fragility, some of which are life-threatening. Yet the evidence base for these remain broadly limited to clinical case studies (Castori et al, 2012; Karthikeyan & Venkat-Raman, 2018; Pezaro et al, 2018). This lack of evidence, knowledge and understanding is not conducive to excellence in maternity care, safer childbearing, or positive experiences in childbirth.

The EDS are a group of multisystemic, inherited conditions that affect connective tissue throughout the body (Malfait et al, 2017). Labelled as the most common systemic inheritable disorder of connective tissue, hypermobile EDS (hEDS) and the related Hypermobility Spectrum Disorders (HSD) are thought to represent 80-90% of EDS cases (Tinkle et al, 2017). Many of those affected experience shared symptoms such as skin hyper extensibility along with joint hypermobility and easy bruising. The focus of this study will be hEDS/HSD, rather than the rarer subtypes of EDS. Whilst contemporary prevalence studies remain absent, in a large

epidemiological study undertaken on a population of 12,853, 3.4% experienced joint hypermobility and chronic widespread pain (Mulvey et al, 2013). These experiences, when used as a proxy for hEDS/HSD resulted in reported estimates of approximately 10 million cases in the United States of America (USA), 19 million in Europe, and 255 million worldwide (Castori et al, 2017; Mulvey et al, 2013). Along with conservative estimates that women are more than twice as likely to be affected than men (2.1:1) (Castori et al, 2014; Demmler et al, 2019), the fact that the world population is comprised of marginally fewer females than males (He et al, 2016), and a crude global birth rate of approximately 130 million per year (United Nations, 2019), the number of births affected by hEDS/HSD can be estimated for the first time here as being approximately 6 million (4.6% of all births) globally per year. For perspective, this number is more than three times higher than the broadly recognised prevalence of intrahepatic cholestasis of pregnancy (0.1-1.5%) (Arrese & Reyes, 2006; Lammert et al, 2000), a condition traditionally afforded far more attention and resources. This new estimate may appear high, but should not be unexpected in a context where recent work has shown a diagnosed prevalence rate of 1 in 500 (Demmler et al, 2019), and only approximately 5% of cases have been successfully diagnosed (Grahame, 2008). This is exacerbated for women where the mean time from the development of significant symptoms to diagnosis is 16 years (EURORDIS AKFF, 2009) and men receive a diagnosis on average 8.5 years earlier than women (Demmler et al, 2019). This lack of timely diagnosis has great implications for pregnancy and birth planning with further research needed to inform the development of services for this population (Demmler et al, 2019).

Patient and public involvement (PPI) and impact data collection activities from a recent review (Pezaro et al, 2018) have identified childbearing as a key transition period for women with hEDS/HSD. Still, whilst qualitative data in relation to women's experiences of hEDS/HSD in the context of motherhood have been researched (De Baets et al, 2017), findings in relation to childbearing remain absent. As such, this is the first study undertaken to arrive at a deeper biopsychosocial understanding of childbearing in the context of living with hEDS/HSD.

METHODS

This research has been undertaken using a narrative approach (Overcash, 2003), underpinned by the interpretive framework of social constructivism (Creswell & Poth, 2017). The reporting of this study is guided by the Standards for Reporting Qualitative Research (SRQR: additional file 1) (O'Brien et al, 2014). The all-female research team comprised a midwife (SP), a Chartered Psychologist (GP) and a General Practitioner (ER) with personal experience of childbearing (SP&ER) and living with hEDS/HSD (GP&ER). The research team had no prior relationship with participants.

Recruitment and sampling

Recruitment took place online via social media, with endorsement from Ehlers-Danlos Support UK, the Ehlers-Danlos Society and the Hypermobility Syndromes Association. Nearly 200 requests to participate were received within 24 hours of recruitment launch. Participants were self-selected. Each needed to meet the following inclusion criteria: English speaking women over the age of 18 who had previously given birth and had a confirmed medical diagnosis of hEDS/HSD or a similar previous diagnosis. Confirmed diagnoses included hEDS as part of the most recent reclassifications (Malfait et al, 2017; Tinkle et al, 2017), along with Ehlers–Danlos

Syndrome Type III according to the Berlin nosology (Beighton et al, 1988), Ehlers–Danlos Syndrome - Hypermobility Type (EDS-HT) in the Villefranche nosology (Beighton et al, 1997), and Joint Hypermobility Syndrome (JHS) as defined by the Brighton Criteria (Grahame et al, 2000), which is recognised to be indistinguishable from EDS-HT (Malfait et al, 2017; Tinkle et al, 2017). Whilst these differing diagnostic labels may appear to imply a heterogeneous group of participants, these terms have all described symptomatic generalised joint hypermobility at different time points. As there remains a lack an objective test or biological marker and criteria continue to evolve for the diagnosis of what is currently referred to as hEDS/HSD, certainty about all of these diagnoses remains elusive at present (see Demmler et al (2019) for further nomenclature information). Individuals with one of the former hypermobile-type diagnoses need not be reassessed against the new criteria and these diagnoses should not be invalidated with the 2017 reclassification (Malfait et al, 2017). As healthcare professionals may come across childbearing women with any of the above diagnostic labels and as management approaches do not differ, we included all historical diagnoses in this work. To ensure that the data remained manageable, the first 40 women to come forward, meet the inclusion criteria and provide their informed consent to participate were invited to contact SP via email to arrange either an online, or telephone-based interview.

Interview procedure

Interviews were completed during May and June of 2018 with SP. Online interviews were completed via Skype software using the synchronous text-based instant messaging method (Pearce et al, 2014). Participants were given access to a dedicated confidential participant Skype account, connected only to the interviewer's professional Skype account.

The interview started with brief demographic questions relating to age, country of origin, number and date of previous births, primary diagnosis and any other diagnoses. Participants were subsequently asked to communicate their narrative following the question "As someone with hEDS [or related diagnosis], we are interested in your childbearing journey. This includes everything from conception to after giving birth. Please may you tell me about your experience?" Follow up questions were also asked to prompt further insights in line with a narrative approach (Stuckey, 2013). Telephone interviews were audio recorded and transcribed verbatim. Data were copied and then deleted from the Skype account following each interview. All transcripts were transferred and saved to a secured electronic folder.

Analysis

Demographic data were analysed using descriptive statistics. Considered to be a complementary approach (Chadwick et al, 2014; Shukla et al, 2014), thematic narrative analysis was used in relation to qualitative data. Using Braun and Clarke's six-phased process inductively (Braun & Clarke, 2006; Clarke & Braun, 2014), analysis focused thematically upon 'what' had been told (Riessman, 2008), with a narrative theoretical lens cast over the data. SP generated initial themes, which were reviewed collectively and refined by the research team (SP, GP & ER).

Patient and Public Involvement

Discussions and engagement via social media highlighted childbearing to be a key transition period for women with hEDS/HSD, where both symptoms and disability can be exacerbated. This led the research team to pursue this topic of enquiry. This open access publication will be accompanied by public engagement and involvement activities as we work with patients,

patient organisations and the public to share these important findings and increase the likelihood of instigating change in practice.

FINDINGS

An international sample of 40 women were interviewed. Of these, 37 were conducted via synchronous text-based instant messaging and three via telephone. Typographical errors have been edited, as this can help readers to read with accuracy and portray participants in the best possible light (Wolcott, 1994). Identities have been anonymised.

Participant characteristics

Participants (age=25-55; year of childbearing=1981-2018; resident country; UK=29 (73%), USA=10 (25%), Canada=1 (2%); diagnosis type = hEDS=9, HSD=0, EDS Type III=11, EDS-HT=16, JHS =4). Of the 52 births reported which did not result in an early pregnancy loss, 38 (70%) were conceived prior to a hEDS/HSD diagnosis, and 16 (30%) were conceived following a diagnosis of hEDS/HSD. Two participants were pregnant with subsequent children at the time of interview. Participants reported several co-morbidities, the most common included Postural Orthostatic Tachycardia syndrome, Irritable Bowel syndrome, asthma, depression and anxiety. See table 1 for a comprehensive overview of demographic data.

----------INSERT TABLE 1 ABOUT HERE----------

Of the 52 births narrated by participants, 15 (29%) were via caesarean section and 37 (71%) occurred vaginally. Labours were clinically induced in 6 cases (12%), whilst in 8 cases (15%), birth was assisted with the use of instruments. The performance of episiotomy was described in

9 cases (17%). Overall, 28 participants (70%) gave birth to one child, 10 (25%) to two children and 2 (5%) had three births.

Thematic narrative analysis

Narrative data are presented in a linear fashion, weaving the events of childbearing in the context of hEDS/HSD together to provide an overarching story (Polinghorne, 1995).

The Antenatal Story

Overall, five themes were derived that describe their experiences in relation to the antenatal period. These related to conception, pregnancy symptoms, knowledge and understanding among health care professionals, poor treatment and decision making.

Conception was relatively easy

Most participants described how conception was "quite easy" (P22) for them. Only a minority of individuals found it difficult to conceive a pregnancy or engaged in fertility treatments.

"My baby was finally conceived after 8 years of trying and after a second attempt at Intracytoplasmic Sperm Injection [ICSI] treatment. She was a frozen embryo for my first and only harvest. I got 29 eggs out (hyper stimulation syndrome)." – P29

Pregnancy symptoms can be exacerbated by hEDS/HSD and vice versa

Once pregnant, participants described a variety of typical pregnancy symptoms such as hyperemesis, pelvic pain, carpal tunnel syndrome and fatigue being particularly challenging for them. They also described how the symptoms of hEDS/HSD such as joint instability and pain can be exacerbated by being pregnant. For example, one woman described having "extra and more frequent pain throughout [her] joints and body that felt more lax than usual" - P14. Others

described dislocations (P25), some whilst sneezing (P28). Another woman (P40) used cannabis to manage her exacerbated hEDS/HSD symptoms. Worsening symptoms seemingly resulted in antenatal admissions and instigated episodes of depression and anxiety in some cases.

Some participants also described complications during pregnancy. These included both high and low risk episodes such as unstable fetal positioning, excess amniotic fluid, haemorrhage, reduced fetal movements and HELLP (Haemolysis, Elevated Liver enzyme levels, and Low Platelet levels) syndrome. Uncertainty in relation to the risks associated with pregnancy and hEDS/HSD was seemingly a cause of anxiety in both healthcare professionals and in the women themselves.

"I was taken off work by the time I was in my 4th month. The hip pain was unbearable. I was working as a nurse in the ICU at that time and by the time I got home I had to crawl to the bathroom at night". - P5

“I had some bleeding at about 6 weeks, which with a history of early miscarriage was a bit disconcerting so I had a couple of early scans. In the end they put it down to me having fragile membranes because of the EDS” - P26

Whilst pregnancy was largely described as challenging, some optimal pregnancy experiences were also reported.

“During pregnancy I felt the healthiest I had been.” – P1

Knowledge and understanding among healthcare professionals

Throughout pregnancy, participants reported how seemingly few healthcare professionals knew how to meet their needs. As an example of this, one participant described how “nobody in the maternity unit had ever heard of EDS so did not know how to manage it” – P24. The need for education, understanding and awareness featured strongly.

"I remember him [junior rheumatologist] saying, you’re not hypermobile because you can’t touch the floor, and I’m thinking I’m eight months pregnant of course I can’t touch the floor." - P30

“I found most of the health care professionals I dealt with were uninformed about EDS. Those who had heard of it would say “ooh the stretchy skin thing” or look at the blue tinge in the whites of my eyes which to be honest is not a true representation of the condition. I don’t personally have the stretchy skin trait for instance. There is a serious lack of knowledge sadly. “– P29

A minority described rare episodes of meeting a professional who understood hEDS/HSD.

“it was refreshing when I saw the anaesthetist and he was aware of EDS. He listened, and it was like a weight had been lifted off me. . . he told me that so many of us with EDS are so used to being not believed.” – P4

Poor treatment

Some women described being treated poorly during pregnancy. Examples of not being listened to or believed featured strongly, along with feeling “hopeless”- P4 and “failed” – P25. One woman explains how she “withdrew from the consultant’s care as [she] couldn’t face the stress

and [her] anxiety was through the roof because the consultant would not listen "- P4. Another felt that she was "treated like a hypochondriac." – P14

"she [consultant] would tell me that I was making up my results as I wanted to hide that I had Gestational Diabetes and that I was making up my diagnosis of EDS as that only makes me good at gymnastics not child birth (that was her line every time I would mention EDS) "– P4

Wanting to be involved in decision making

This sample of women expressed a desire to be involved in decision making about their care, particularly in relation to mode of birth. However, their involvement in effective decision making was often hindered by a lack of evidence-based conversations or "tailored advice" – P34.

"I wanted desperately to have a conversation about the risks of vaginal vs caesarean birth for somebody in my position but was told that nothing about a birth plan would be decided until 6 weeks before. And at the appointment, I was merely asked if I'd decided which I wanted." – P34

"I had expressed to my doctor that I wanted to have a C-section only because I have seen some really bad deliveries before. "– P28

"Fortunately, they were all happy to support the advice given and help me plan. I had an elective section in the end. "– P21

The intra-partum story

Three themes were derived in relation to the intra-partum period. These related to the durations of labour and birth, ineffective medicines and birth trauma.

Labours can be long, but births can be fast

The World Health Organisation categorises the latent phase of labour by painful uterine contractions and variable changes of the cervix, including some degree of effacement and slower progression of dilatation up to 5 cm for first and subsequent labours (World Health Organisation, 2018a). Active labour is categorised by regular, painful uterine contractions, a substantial degree of cervical effacement and more rapid cervical dilatation from 5 cm until full dilatation for first and subsequent labours (World Health Organisation, 2018a). The build up to active labour was typically described by these women as being "slow" – P27. As such, some women described being in the latent phase of labour for "weeks" – P15, and some were told they had an "irritable uterus" – P13. Whereas active labours and births were typically described as being "very very fast" – P30. A precipitate, or abnormally rapid labour is clinically defined as the expulsion of the baby within less than 3 hours of commencement of regular contractions (Suzuki, 2015). For maternity staff this occurrence was seemingly an unexpected "shock" – P16.

The following two participant quotes come from two women, each birthing for the first time.

"At 7am I was examined and was 1cm, at 7:30am I asked my husband to get the midwife as 'it was happening' she examined me again and I was 4cm. We went to the delivery suite, and after a little bounding on a ball, and some gas and air, with 3 big pushes I gave birth at 9:30am." – P9

"I was advised that I had now reached 10cm - fully dilated - which was just 1.5 hours after the 1-2 cm examination." – P16

The drugs don't always work

In line with current understandings (Hakim et al, 2005), some participants experienced poor regional anaesthetic and analgesic coverage along with adverse drug reactions. Yet here, these experiences were often confounded by verbal reports of pain being ignored. In such cases some participants described how staff "operated anyway"– P1.

"I can still vividly remember the feeling of the thread [suturing] being pulled through my skin and feel that I'm still a bit traumatised by this". – P3

"I was in agony, I could feel burning and ripping sensations. They tried topping up my epidural, but nothing worked." – P2

Birth Trauma

When describing the births of their babies in a context of having a hEDS/HSD diagnosis, many participants report episodes of both physical and/or psychological trauma. Participant 20 described childbearing with hEDS/HSD as "quite a traumatic experience all around, from start to finish", leading her to decide only to "have one child". When obstetric complications arise, participants also described themselves, their partners and clinicians as being "panicked" - P4. Monitoring women with hEDS/HSD in high risk situations can also be seemingly problematic due to their skin being "loose" – P34. Episodes of birth trauma were reportedly exacerbated by poor care, substandard facilities and not being "taken seriously" – P2. Babies also experienced physical birth traumas in three reported cases.

"My husband and I walked away feeling trauma from our experience given that we prepared for work that day, went to a medical appointment figuring it was routine… And shortly after I was

told that I might lose both of the babies and I was going into spontaneous labour. It was shocking and unexpected" -P15

"My husband looked at me in full horror" [baby was rushed out of the room not breathing as the mother was haemorrhaging]. – P36

The postnatal story

Five themes were identified in relation to how this group of women experienced the postnatal aspects of childbearing in this context. These include reflecting on the presence or absence of a diagnosis throughout the childbearing journey, struggling to recover, looking back in anger, the challenges of motherhood and the fact that babies suffer too.

Reflecting on the presence or absence of a diagnosis throughout the childbearing journey

Participants often reflect on how the presence or absence of a diagnosis affected their childbearing experiences overall. Some regretted that both they and healthcare professionals had not been more aware and understanding of their condition in hindsight.

"If I'd been diagnosed before pregnancy there are things I would not have even contemplated. Water birth, birthing centre for first birth - I would have also researched pain relief etc. "– P39

"I have heard that some women with EDS are advised to have a C-section, so I am glad that I didn't know back then." – P16

Struggling to recover

Often, women described their struggle to recover from birth, both physically and psychologically. Narratives in relation to pain, fatigue and poor wound healing featured

strongly. In some cases, “wounds reopened “- P39. A straightforward postnatal recovery was described in only a minority of cases.

“I had from my son an episiotomy and I now still have problems with prolapse, but yeah you don’t know really how to compare it to other people, but certainly pain afterwards was still there with the healing, and it seemed to take quite a long time to heal. I think the prolapse is maybe part of the elasticity and the whole problems and the bowel side as well, I still have problems there.” -P38

“It feels like I may have put something out of place in the coccyx area. Even now when every time I take my underwear up and down it’s a really sharp pain in the tissue. Worst thing is trying to maintain a decent position for feeding especially at night. I'll end up sliding down and trying to shift my weight onto one side because it's agony. But then that puts strain on my collar bone which tends to be adrift or pop a rib out or something “– P34

Looking back in anger

Several participants ruminated with frustration. Some were angered by a lack of support and delays in treatment. Others reflected with anger about their poor experiences within maternity services.

“If I buzzed for help because I couldn't sit up to get [my baby] I was made to feel like a failure. I didn't breastfeed because I was told if I did my hormones would carry on and my hips would still be as bad, but I was made to feel bad for that decision. “– P2

“I was told I was lazy for not changing my son, the truth was - I was in agony and struggled to get the help I desperately needed. I felt I had to push through it, and it had a detrimental effect both mentally and physically. “- P14

Motherhood poses new challenges

Participants encountered new challenges in motherhood. Such challenges included difficulties in holding, caring for, bonding with and breastfeeding their babies. In some cases, this resulted in repeated joint dislocations. The need for additional support featured strongly.

“I have lots of pain and subluxation in my wrists, I suffer from back pain from lifting or holding her. Every day is a constant struggle to cope”. – P33

“Trying to do compressions and maintain a good breast shape [when breastfeeding] while holding the baby are all extremely painful and difficult. I think there could be more awareness in delivery aftercare staff with regards to positioning for feeding for people with disabilities.” – P34

Babies suffer too

Participant 10 described childbearing as being a “shitty time” for her son with “post birth interventions” and “reflux”. Other babies displayed symptoms of joint instability and went on to receive a confirmed diagnosis of hEDS/HSD. The bruising found on one baby led to a mother being suspected of dishonesty.

“My son, at almost age 9, is displaying some of the signs of also having hEDS. Namely the Beighton elbows, knees, and long slender fingers that wrap around the wrists. “– P39

"On my 3 day review a midwife came to the house, she started questioning me about how [my baby] got her bruises, she had read c section but didn't read the rest of the notes. When I explained to her, she said they never use forceps in c section, went back and read the notes and then apologised." – P2

DISCUSSION

This article is the first to highlight that an estimated 6 million childbearing women per year are affected by hEDS/HSD globally, meaning that cases of hEDS/HSD should no longer be considered rare in maternity services. Yet, in the current study, maternity staff were observed to be panicked due to a lack of knowledge and understanding in delivering maternity care in this context, where local and regional anaesthetics were reportedly less effective and long latent phases of labour quickly developed into rapidly progressing active labours and births. The postnatal phase was also impacted with challenges including difficulties in holding, caring for, bonding with and breastfeeding babies. This need for additional support featured strongly and aligns with infant feeding experiences of women with autoimmune rheumatic diseases (Williams et al, 2019).

For these women, the symptoms and complications associated with hEDS/HSD, motherhood and childbearing were broadly exacerbated. For some, poor experiences led to trauma, stress, anxiety, disengagement and decisions to avoid future childbearing. The findings presented here corroborate those of De Baets and colleagues (2017), where women with EDS-HT also often chose not to have any further children as a result of their negative experiences. However, whilst previous research has reported higher levels of infertility within EDS populations (Hurst et al,

2014), many participants here described conception as relatively easily. Despite this, inferences cannot be made about fertility, given that the inclusion criteria were set to include those with experience of childbearing. Additionally, whilst the findings presented here are quite subjective due to the selected study design, seemingly shared decision making along with the amount of respect and support given by caregivers and the quality of the caregiver-patient relationship appear to be as important in childbearing for this group as they are for the general birthing population (Bell and Andersson, 2016; Cook and Loomis, 2012; Hodnett, 2002).

A key strength of this study is that it is the first of its kind to offer a voice to women with hEDS/HSD in relation to their childbearing experiences. The methodology used has also enabled these participants, some with debilitating symptoms, to engage in research. Furthermore, the receipt of nearly 200 requests to participate within 24-hours of recruitment launch indicates a high desire for research in this field.

Nevertheless, whilst we do not doubt the validity of these women's accounts, they have not been verified alongside medical records or the new 2017 criteria (Malfait et al, 2017), and we are unable to establish the causation of outcomes, which may have also been compounded by the co-morbidities reported. Moreover, new hEDS/HSD diagnostic criteria are still under discussion (Tinkle et al, 2017). Additionally, the figure used to estimate universal prevalence rates in maternity services was calculated using data from a UK population who experienced joint hypermobility and chronic widespread pain, used as a proxy for hEDS/HSD, rather than those with a confirmed diagnosis (Mulvey et al, 2013). More robust estimates are needed.

The concerns of childbearing women with hEDS/HSD must be given credence. Clinicians and policy makers may consider the implementation of further clinical training, guidance and education in this area to improve the quality, safety and experience of maternity care. Larger quantitative studies would also be useful, along with studies investigating the perspectives of wider family members. Moreover, it will be important to gather the perspectives of maternity staff delivering care to childbearing women with hEDS/HSD and their babies. Crucially, new research can now investigate whether symptoms previously considered to be typical of pregnancy may actually be attributed to hEDS/HSD due to the new higher estimates presented.

CONCLUSIONS

Prior to this study, cases of hypermobile Ehlers-Danlos syndrome (hEDS) and the related Hypermobility Spectrum Disorders (HSD) were believed to be rare in maternity services. Cases of hEDS/HSD in the context of childbearing are associated with a number of life-threatening obstetric complications and morbidities. This study presents new estimated prevalence rates which suggest that hEDS/HSD should no longer be considered to be rare in maternity services. For childbearing women with hEDS/HSD, a poor experience of maternity care can lead to a disengagement from services, trauma, stress, anxiety and an avoidance of future childbearing. Maternity staff were perceived by childbearing women with hEDS/HSD to be panicked and lacking in knowledge and awareness when delivering maternity care to them. Further training, guidance and education may be required to prepare maternity staff for this new reality. As a first step, www.hEDSTogether.com has been developed to provide a repository of evidence in relation to this topic, along with a freely downloadable toolkit for use in practice.

List of abbreviations

Ehlers-Danlos Syndromes (EDS)

Hypermobile EDS (hEDS)

Ehlers-Danlos syndrome Type III (EDS Type III)

Ehlers-Danlos syndrome – Hypermobility type (EDS – HT)

Hypermobility Spectrum Disorders (HSD)

Hypermobility Syndromes Association (HMSA)

The Ehlers-Danlos Support United Kingdom (EDS-UK)

Joint Hypermobility Syndrome (JHS)

The (UK) National Health Service (NHS)

Patient and Public Involvement (PPI)

Standards for Reporting Qualitative Research (SRQR)

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Table 1: Demographic analysis of participants

Participant Number	Age	Country of origin	Year birth(s) occurred	Number of biological children	Year of EDS diagnosis	Type of diagnosis	Other health Conditions (Year of Diagnosis according to participant)
1	52	UK	1993 1998	2	2006	EDS - HT	• IBS (1985) • Asthma (1985) • Fibromyalgia (2007) • ME/CFS (2007) • POTS (2009)
2	28	UK	2012	1	2017	hEDS	• Fibromyalgia (2014) • Occipital neuralgia (2016) • POTS (2017)
3	35	UK	2008	1	2010	JHS	• IBS (2004) • Osteoarthritis (2010)
4	39	UK	2015	1+ pregnant	1999	EDS - HT	• IBS (1999) • Anxiety (1997)
5	37	USA	2005 2010	2	2008	EDS - HT	• Migraine (Since childhood) • Dissected left carotid artery (2012) • Hypothyroid (2014)
6	49	UK	1997 2003	2	2014	EDS - HT	• Fibrositis (as a teenager) • Endometriosis (2001) • Mild scoliosis (2005) • CTS (2012) • Chronic migraine with associated neurological features (2014) • Extra renal pelvis and duplex left Kidney (2016). • Irritable bladder (ongoing).
7	34	USA	2007 2012	2	2016	EDS - HT	• Osteolysis distal clavicle (2001) • Arthritis (2001) • Herniated disc L5/S1 (2002-4) • Discectomy and laminectomy resulting in arthrosclerosis (2002) • Artificial disc replacement (2004) • HELLP Syndrome (2007) • PIH (2012) • Obstructed kidney stone resulting in sepsis (2015)

							• TMJdisorder (2016) • Symptomatic Premature ventricular complexes (2016) • POTS (2016) • MCAS (2017) • Spontaneous CSF leak (2017) • Paradoxical vocal fold motion (2017) • Obstructive sleep apnoea (2018) • Sepsis (2018)
8	45	UK	2009	1	2018	hEDS	• Endometriosis (1998) • Chronic atypical migraine (2011) • Formaldehyde allergy and chronic allergic eye disease in (2013) • Fibromyalgia (2013) • Chronic fatigue (2013) • TMJ dysfunction (2011) • Recurring anaemia (since childhood)
9	25	UK	2017	1	2017	hEDS	• Allergic rhinitis (2002) • Asthma (2015) • PGP (2016) • Iron deficiency (2017) • POTS (2017)
10	39	UK	2011 2014	2	2015	EDS - HT	• Drug induced lupus (2001-02) • PCOS with insulin resistance (2006) • Coeliac disease (2009) • Hypothyroid (2014) • Perennial rhinitis
11	35	UK	2001 2008 2014	2 + still birth in 2001	2017	hEDS	• PCOS (2002) • POTS (2014) • Floppy bladder (2015) • Severe bradycardia (2017)
12	40	UK	2004	1	2009	EDS - HT	• Asthma (1980) • Toxoplasmosis (1988) • POTS (2014)
13	34	USA	2009	1	2018	hEDS	• Asthma (1991) • Patellofemoral Chondromalacia (2007) • Fibromyalgia (2016)

14	31	UK	2007	1	2017	hEDS	• Irlens Syndrome (unknown) • General anxiety and depression (2000) • Autism (2016) • ADHD (2016) • Dyslexia (2016) • Dyspraxia (2016) • Gastroparesis, wheat intolerance and dietary issues including severe IBS type symptoms (2016) • Severe hearing loss (2018)
15	31	USA	2014	2 (Twins)	2015	EDS - HT	• Asthma (Since childhood) • Fibromyalgia (2005) • Gastritis (2013) • Chiari malformation and tethered cord syndrome (2015) • Dysautonomia (2015) • POTS (2015)
16	44	UK	2004	1	2014	EDS - HT	• Recurrent patellar dislocations (1978) • Tonsillectomy and adenoidectomy (1980) • Seasonal asthma (1990) • Cervical erosions (since early adulthood) • PCOS (2008) • Uterine polyp (2011) • Marfanoid Habitus (2014)
17	28	UK	2018	1 + 1 miscarriage	2011	EDS Type III	• Stress & exercise induced asthma (2002) • IBS (2005) • PCOS (2006) • Johanson-Larson disease (osteochondrosis) (2006)
18	55	Canada	1981 1983	2	2017	hEDS	• Hypertension (2017)
19	30	UK	2017	1	2012	EDS Type III	• Lumbago (2002) • Chronic migraines (2008) • POTS (2012) • CFS (2012) • Osteopenia (unknown) • Raynaud syndrome (unknown)
20	43	USA	2002	1	2015	JHS	• Eczema (1978)

							• Asthma (1980) • Allergies (1980) • Arthritis (1990) • Undifferentiated connective tissue disorder (1998) • Migraines (2002) • Post-Partum Depression (2002) • Rotator cuff tendonitis (2004) • Gluten intolerance (2005) • Eosinophilic esophagitis (2009) • Gastroesophageal reflux disease (GERD) (2009) • Hip bursitis (2010) • Chronic Ethmoidal Sinusitis (2017) • Patellar tendonitis (2017)
21	39	UK	2009	1	2009	JHS	• Postural Hypotension (1997) • Benign Sinus Tachycardia (2002) • POTS (2008) • IBS (unknown) • Migraine (unknown)
22	55	UK	1991 1994	2	2015	EDS Type III	• Chronic fatigue syndrome (2014) • Fibromyalgia (2014) • Autism (2016) • Arthritis (unknown)
23	32	USA	2017	1	2017	EDS - HT	• Achalasia (2010)
24	28	UK	2018	1	2015	EDS - HT	• Fibromyalgia (2016)
25	38	USA	2010	0 (Stillbirth)	2011	EDS - HT	• Fibromyalgia (unknown) • Chronic migraine (unknown) • Osteoarthritis in hips (unknown) • Degenerative disc disease (unknown) • Scoliosis (unknown) • Kyphosis (unknown) • Sciatica (unknown) • Spondylitis (unknown) • Rhinitis (unknown) • Sinusitis (unknown) • Anxiety (unknown)

							• Depression (unknown) • Flat feet/foot problems (unknown) • IBS (unknown) • kEDS (unknown) • Tendonitis in elbows and feet (unknown) • PTSD (unknown) • Tarlov Cysts (unknown)
26	30	UK	2016	1 + 1 Miscarriage	2014	EDS Type III	• Asthma (As a child) • Anorexia nervosa (2007) • POTS (2009) • Mitral valve prolapse (2009) • Reflex anoxic seizures (2009) • Mild gastroparesis (2011) • Reactive hypoglycaemia (2012) • Post-natal depression (2016)
27	34	UK	2015	1	2012	EDS - HT	• Asthma (from childhood) • Anxiety (from childhood) • POTS (2012)
28	35	USA	2011	1+ 1 Miscarriage	2018	hEDS	• Tonsil and adenoid surgery (1989) • Chronic pain (Since 1994) • Partial hysterectomy and scar tissue removal (2002) • Cervical cerclage (2011) • Fibromyalgia (2018)
29	35	UK	2017	1	2015	EDS Type III	• Asthma (1995) • Endometriosis (2009) • Fibromyalgia (2012) • Spine problems (2012) • Pigmented villonodular synovitis (diffuse type) (2013) • Vertigo (2013) • Allergies (2014)
30	38	UK	2003 2005 2009	3	2006	EDS Type III	• Migraines (2000) • Recurring depression (2000)
31	34	UK	2016	1 + 1 Miscarriage	2012	JHS	• Asthma (1996)
32	33	USA	2016	1	2002	EDS Type	• Depression (2012)

						III	• Anxiety (2012) • Preeclampsia (2016) • PCOS (2018)
33	30	UK	2017	1	2017	hEDS	• Chronic migraine (2002) • Mannose binding lectin deficiency (2012) • POTS with vasopressive autonomically mediated syncope (2016)
34	41	UK	2018	1	2010	EDS Type III with benign JHS	• Hypertension (2003) • Insulin resistance (2004)
35	32	UK	2017	1	2016	EDS - HT	• Incomplete Marfan (unknown) • MCAS (1999/2000) • Gall stones (2004) • PCOS (2015) • Meibomian gland dysfunction (2018)
36	29	USA	2013	1 + 3 Miscarriages	2018	EDS - HT	• Chronic Meningitis (unknown) • Scoliosis (unknown) • Ligament/tendon/muscle ruptures (unknown) • Rhabdomyolysis (unknown) • Lupus (unknown) • Lumbar Spine herniations (2013) • Spine cyst (unknown) • Arachnoiditis (unknown) • Complex regional pain syndrome (unknown) • Spondylolisthesis (unknown) • Sacroiliac joint dysfunction (unknown) • Nerve entrapment (unknown) • POTS (unknown) • CSF leak (unknown) • Neutropenia/leukopenia (unknown) • Hypertensive episodes (unknown) • Syncope (unknown) • Radiculopathy (unknown) • Neuropathy (unknown) • Roseola (unknown) • Reoccurring thoracic outlet syndrome (2018)

37	30	UK	2014	1 + Current pregnancy	2014	EDS Type III	• Mild asthma (2008) • Anxiety (2011) • Depression (2011)
38	47	UK	2001	2 (twins) +2 miscarriages	2008	EDS Type III	• Pre-eclampsia (2001) • Kidney failure (2001) • Twin to twin transfusion (2001)
39	37	UK	2007 2006	2	2018	Benign JHS EDS - HT	• Scoliosis & spondylolysis (2002) • Coagulase- negative staphylococci (2011) • ME (2016) • Chronic migraine (2016) • Hiatus Hernia (2016) • Anaemia (2018) • POTS (2018) • Spinal arthritis (2018)
40	42	UK	2015	1	2004	EDS Type III	• Autonomic dysfunction (2006)

Table 1 abbreviations: Myalgic encephalomyelitis or encephalopathy (ME); Chronic Fatigue Syndrome (CFS); Irritable Bowel Syndrome (IBS); Postural Orthostatic Tachycardia Syndrome (POTS); Carpal Tunnel Syndrome (CTS); Pregnancy induced hypertension (PIH); Haemolysis, Elevated Liver enzyme levels, and Low Platelet levels (HELLP); Temporomandibular joint (TMJ); Mast cell activation syndrome (MCAS); Cerebro Spinal Fluid (CSF); Temporomandibular joint dysfunction (TMJ); Pelvic Girdle Pain (PGP); Polycystic ovary syndrome (PCOS); Attention deficit hyperactivity disorder (ADHD); Kyphoscoliotic Ehlers-Danlos syndrome (kEDS); Post-Traumatic Stress Disorder (PTSD).