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The Impact of Pain on Everyday Activities of People With Hypermobility Spectrum Disorders or Hypermobility Ehlers Danlos Syndrome

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ABSTRACT

Background: This study describes aspects of pain and how pain affects everyday life and examines the relation between chronic pain and activity limitations in people with hypermobility spectrum disorders (HSD) or hypermobility Ehlers Danlos syndrome (hEDS).

Methods: This cross-sectional study used data from 2016 to 2021 obtained from the Swedish quality registry for pain rehabilitation (SQRP), comparing those with HSD/hEDS with the larger group of people with mixed chronic pain conditions as a reference group (RG).

Results: Of the 43,801 people registered in the SQRP, 1211 (2.8%) were diagnosed with HSD/hEDS (88.9% women). The mean age of the HSD/hEDS group was younger (36.3 ± 11.8) than the RG (45.7 ± 12.8). The HSD/hEDS group had a statistically significant ($p < 0.001$) earlier onset of pain (calculated in years) in contrast to the RG. In the HSD/hEDS group, 80.1% had persistent pain; in the RG, 74.2% had persistent pain. The HSD/hEDS group reported more pain locations (20.0 ± 7.9) than the RG (14.8 ± 8.8). The HSD/hEDS group reported more problems performing leisure, social, and household activities than the RG; however, pain intensity was statistically significantly lower ($p < 0.001$) in the HSD/hEDS related to the RG.

Conclusions: There were indications that pain affected daily activities for people with HSD/hEDS, who had earlier onset of pain, marked more pain locations, and had more persistent pain, but pain intensity was not as decisive in contrast to the RG.

Significance Statement: In a comparison yielding statistically significant results ($p < 0.001$), persons with hypermobility spectrum disorder (HSD) or hypermobility Ehlers-Danlos syndrome (hEDS) reported earlier pain onset, longer pain durations, and a greater number of pain locations but surprisingly, lower pain intensity than the reference group which consisted of a mixed group of pain conditions. These pain characteristics affected daily activities, indicating a substantial impact on daily life for those with HSD/hEDS.

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1 | Introduction

It is still quite unknown how extensive and widespread pain is among people with hypermobility diagnoses, whether specific areas of the body are affected more often, or how pain affects the activities of everyday life (Chopra et al. 2017; Kalisch et al. 2020). Hypermobility, defined as mobility beyond the normal range of motion in a joint (Castori et al. 2017; Malfait et al. 2017) has an estimated prevalence in adults of 10%–15% with the majority being women (Castori et al. 2010; Tinkle et al. 2017). The most common hypermobility diagnoses are Hypermobility spectrum disorder (HSD) and Ehlers-Danlos syndrome (EDS), where the hypermobility variant (hEDS) is clearly dominant (80%–90%) of the 13 different EDS variants. HSD and hEDS are hereditary and congenital (Castori et al. 2017; Tinkle et al. 2017). HSD and hEDS are based on diagnoses criteria from 2017 (Malfait et al. 2017). Typical characteristics in HSD/hEDS include local or general hypermobility, which can lead to joint instability and musculoskeletal problems such as pain and tendinopathies (Castori et al. 2017; Malfait et al. 2017).

People with HSD/hEDS experience pain more often than the general population; previous studies show that chronic pain occurs close to 90% of cases (Chopra et al. 2017; Voermans et al. 2010). The pain process usually begins with local or regional pain, often associated with joint trauma as the result of, for example, strain or dislocation. Over time, the nociceptive pain increases the sensitivity of the somatosensory system; thus, a wide-spread nociplastic pain develops, aggravated by new peripheral input, and negatively impact activity and function in daily life and cause difficulties performing activities (Chopra et al. 2017; Malfait et al. 2021; Syx et al. 2017). Over 95% of individuals diagnosed with hypermobility in pain clinics were found to have hEDS (Nicholson et al. 2022). Previous research (Ritelli et al. 2024) as well as clinical experience demonstrate the need for studies that can differentiate HSD/hEDS from other pain conditions. This differentiation is crucial as care and rehabilitation must be tailored to the specific impacts of hypermobility on daily life.

People with HSD/hEDS have described recurring visits to healthcare professionals for various symptoms such as pain or repeated sprains, but obtaining the correct diagnosis can take years despite recent advancements in research and clinical tools such as diagnostic checklists (Kalisch et al. 2020). Since the symptoms often start at a young age and the societal costs are significant, an accurate and rapid diagnosis is valuable. Distinguishing their specific characteristics enables optimal treatment and tailored rehabilitation, giving people with HSD/hEDS a chance for an active and independent life, reduced suffering, and reduced care utilisation.

This study aims to investigate how pain is expressed in people with HSD/hEDS, using a larger group with mixed chronic pain conditions as a point of reference. The specific objectives are to (1) describe pain expressions as pain intensity, pain location, and pain duration, identifying distinctive patterns and (2) to examine the relation between chronic pain and activity limitations in people with HSD/hEDS.

2 | Methods

2.1 | Settings

This cross-sectional study used data from the Swedish quality registry for pain rehabilitation (SQRP). Most specialist pain clinics (90%) in Sweden collect data for the SQRP from patients ≥ 18 years old with complex chronic pain, from baseline and after completing the interdisciplinary pain rehabilitation programme (IPRP) (Fischer et al. 2020). This study used baseline data (i.e., at time for the assessment before IPRP). Patients were informed that completed questionnaires could be used for research purposes at the time of inclusion in the SQRP. A written consent form had also been confirmed by the patient. This study has followed ethical guidelines and regulations according to the declaration of Helsinki (World Medical Association 2013) and received approval from the Swedish ethics review authority (DNR: 2021-05775-01). We used data from selected patient-reported outcome measures (PROMs) reflecting how pain affects everyday life, collected from the SQRP between August 2016 and December 2021.

Inclusion criteria for persons with hypermobility diagnoses were based on international classification diseases (ICD)-10-SE with diagnosis codes M35.7 hypermobility spectrum disorder (HSD) and Q79.6 Ehlers Danlos syndrome (EDS). In ICD-10-SE as well as in the SQRP, all 13 Ehlers Danlos diagnosis were recorded as one group: Q79.6. Exclusion criteria are as follows: if a person had contact with the pain clinic in several treatment occasions, the first occasion was counted; that is, a person could only be included once. The HSD/hEDS group consisted of 1211 persons of the 43,801 unique persons in the SQRP.

Patients included in the SQRP are typically referred from primary care to specialist pain clinics. However, the referral process is heterogeneous and may also originate from other specialist clinics. The patient may have received a previous diagnosis of HSD/hEDS, or this diagnosis may be made by the pain physician during the initial visit to the pain clinic.

Corresponding data were obtained for the reference group (RG) from the SQRP. The RG consisted of a mixed group of pain-related diagnoses, including fibromyalgia, lumbago, and myalgia and nociceptive, neuropathic, idiopathic, and unspecified chronic pain. The purpose of including the RG in the present study was to serve as a point of reference to highlight potential differences between HSD/hEDS and typical pain rehabilitation patients. The exclusion criteria were the same for the RG as they were for the HSD/hEDS group; that is, the person could only be included once. The RG consisted of 42,590 persons.

2.2 | Health Information and Sociodemographic Data

All data derive from the SQRP. Health information (Table 1) comprise sex, age, body mass index (BMI), highest level of completed medical visits, and number of medical visits in the past year due

TABLE 1 | Demographic characteristics from HSD/hEDS and the RG.

	HSD/hEDS *N* = 1211		RG *N* = 42,590		
	M (SD)	%	*M* (SD)	%	*p*
Age (years)	36.3 (11.8)		45.7 (12.8)		< 0.001
18–39		62.3		31.5	
40–65		36.4		62.9	
> 66		0.7		5.3	
Sex					
Women		88.9		74.3	
Men		11.1		25.7	
BMI	26.3 (6.5)		27.2 (6.7)		< 0.001
Highest school education (completed)					
Lower secondary school		10.2		9.9	
Higher secondary school		43.3		36.3	
University education		32.0		29.8	
Work/studies/work training		65.2		57.8	
Compensation from the insurance fund		40.4		45.1	
< 50%		9.7		11.1	
> 50%		29.8		32.8	
Doctor visits in the last year due to pain					
0–3		42.2		33.6	
> 4		54.9		61.9	
Exercise/week					
None or < 30 min		15.6		17.2	
30–90 min		29.4		30.1	
90–300 min		34.3		32.1	
> 300 min		18.2		15.8	

Abbreviations: BMI, body mass index; Exercise/week, exercising, e.g., walking, cycling, or gardening, an average week (minutes); hEDS, hypermobility Ehlers Danlos syndrome; HSD, hypermobility spectrum disorders; *M*, mean; *N*, numbers; RG, reference group; SD, standard deviation; %, percent.

to pain from the SQRP. Sociodemographic data comprise work, studies, and work training (percentage [%] of how much time the people spent working or engaged in educational pursuits) and questions about support or compensation from the Swedish social insurance agency due to illness (yes/no and if yes what percentage [%] of full time) (Table 1). Additionally, we used information about the amount of time spent on daily exercise based on a 7-point scale, in this study re-categorised into four categories (from 0 to < 30 min, 30–90 min, 90–300 min, and > 300 min). Data were also collected about the onset of pain (days) and whether the pain was persistent (year, month, and date of onset) or recurrent.

2.3 | Outcome Measures

The selected PROMs comprise (Table 2) the 11-point numeric pain rating scale (NPRS) indicating the average pain intensity experienced during the last week from 0 (no pain) to 10 (worst possible pain) (Dworkin et al. 2008), the pain region inventory (PRI), a form with 36 predefined locations, 18 on the front and 18 on the back of the body indicating the placement and dissemination of the pain (Gerdle et al. 2021): (1) head/face, (2) neck, (3) shoulder, (4) upper arm, (5) elbow, (6) forearm, (7) hand, (8) anterior chest, (9) lateral side of chest, (10) belly, (11) sexual organs, (12) upper back, (13) low back, (14) hip/gluteal area, (15) thigh, (16) knee, (17) shank, and (18) foot (Figure 1 and Table S2). The multidimensional pain inventory Swedish version (MPI-S) evaluates psychosocial and behavioural consequences of chronic pain (Kerns et al. 1985). The instrument consists of 34 questions in two parts. The present study used part I, which focuses on interference in everyday life (11 questions) (Tables 2, 3 and Table S1). The score ranges between 0 and 6 where a low value indicates lower intensity (Bergström et al. 1998).

TABLE 2 | Pain characteristics from the SQRP for HSD/hEDS and RG.

	HSD/hEDS $N=1211$		RG $N=42{,}590$		Welch's *t*-test (*p*)	Mann–Whitney *U* (*p*)
	M **(SD)**	***N*** **(%)**	***M*** **(SD)**	***N*** **(%)**		
Pain intensity during the last week (NPRS [0–10])	6.4 (1.8)		6.8 (2.0)		<0.001	
Onset pain (years)	13.5 (10.9)		9.5 (9.8)		<0.001	
Years with persistent pain	10.4 (9.5)		7.6 (8.6)		<0.001	
Number of pain locations	20.0 (7.9)		14.8 (8.8)		<0.001	
Is your pain periodically recurring or persistent yes/no?		978 (80.8)		31,618 (74.2)		0.017

Abbreviations: hEDS, hypermobility Ehlers Danlos syndrome; HSD, hypermobility spectrum disorders; *M*, mean; *N*, numbers; NPRS 0–10, numerical pain rating scale (pain intensity during the last week); Persistent pain and Recurring pain, question from health information "Is your pain periodically recurring or persistent yes/no?"; PRI 0–36, pain region index (number of pain locations); RG, reference group; SD, standard deviation; %, percent.

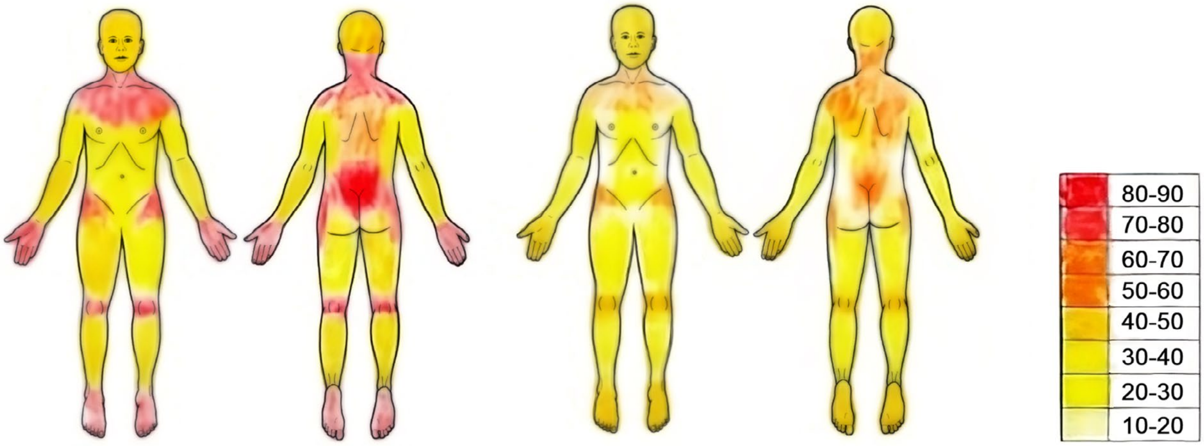

FIGURE 1 | Colour gradient from white (no pain) to red (greatest pain) based on the number of pain locations (in %) marked in PRI-index, HSD/hEDS (left image), and RG (right image).

2.4 | Statistical Analyses

International business machines corporation (IBM) statistical package for the social sciences (SPSS) statistics (version 29.0; IBM Corporation, Armonk, NY: IBM Corp, USA) (Corp 2022) was used for statistical analysis. The descriptive statistics (Tables 1, 2, and 3) used percentages (%) for categorical variables or when more than one answer option was available for the same question, such as for subscales in the MPI-S regarding questions about interference with daily activities (Table 3). Welch's *t*-test and the Mann–Whitney *U* test were used to examine differences between the HSD/EDS group and the RG. To calculate the effect size (r) in the Mann–Whitney-*U* test, we converted the test statistic to a z-score, which was divided by the square root of numbers ($\sqrt{n}$). A critical alpha level of 0.001 was deemed to be a reasonable compromise for balancing Type I and Type II errors.

We used linear regression models to determine the influence of pain variables on outcomes (Tables 4 and 5). The self-reported pain intensity during the last week (NRPS 0–10) was used as the dependent variable. We tested whether there was an association between following independent variables: number of pain locations (PRI-index), persistent pain, and intermittent pain. Furthermore, interference in daily activities was analysed using the MPI-S subscales as the dependent variable; we tested whether there was a relationship between the following independent variables: pain duration, number of pain locations, and pain intensity during the last week (NRPS, 0–10).

3 | Results

There were 43,801 unique people registered in the SQRP, of which 2.8% were diagnosed with HSD/hEDS (Table 1). The HSD/hEDS group was almost a decade younger ($M=36.3$, $SD=11.8$) than the RG ($M=45.7$, $SD=12.8$). The HSD/hEDS group also had a higher proportion of women (88.9%) than the RG (74.3%).

More people in the HSD/hEDS group than the RG were working, engaged in work training, or engaged in studies (HSD/hEDS = 65.2%, RG = 57.8%). In the HSD/hEDS group, fewer people received compensation from the social insurance agency (HSD/hEDS = 40.4%, RG = 45.1%) and they reported fewer visits to the physician in the past year due to pain, unlike the RG. Both groups exercised daily at an equivalent level, with a median (MD) of 5 and an interquartile range of 3–6 (Table 1). There

TABLE 3 | Interference in daily activities in HSD/hEDS and RG.

	HSD/hEDS $N=1211$		**RG $N=42{,}590$**		**Welch's t-test (p)**	**Mann–Whitney U (p)**
	***M* (SD)**	**Md [IQR]**	***M* (SD)**	**Md [IQR]**		
Interference MPI-S	4.2 (1.2)		4.3 (1.2)		< 0.001	
Interference in daily activities		4.8 [3.7–5.7]		5.0 [3.9–5.8]		0.004
Work ability		5.3 [4.1–6.0]		5.4 [4.4–6.0]		0.009
Leisure activities/social life		5.2 [4.0–5.9]		5.2 [4.2–5.9]		0.021
Participate in activities		4.7 [3.4–5.6]		4.8 [3.6–5.7]		< 0.001
Activity restriction		4.4 [3.1–5.3]		4.6 [3.3–5.6]		< 0.001
Enjoyment with family/friends		4.3 [3.0–5.4]		4.6 [3.3–5.6]		< 0.001
Changed relationships		3.4 [1.6–4.8]		3.6 [1.9–5.0]		< 0.001
Impact on job satisfaction		4.2 [2.9–5.4]		4.5 [3.2–5.6]		< 0.001
Ability to do household chores		4.4 [3.1–5.5]		4.4 [3.1–5.5]		0.271
Ability to plan		4.3 [2.6–5.5]		4.5 [3.0–5.6]		0.002
Relation to friend/acquaintance		3.9 [2.0–5.2]		4.0 [2.3–5.3]		0.014

Note: Questions in questionnaire MPI-S Interference: Interference in daily activities (Interference in daily activities), Changed ability to work since onset of pain (Work ability), Change regarding the area of leisure activities/social life (Leisure activities/social life), Ability to participate in leisure and social activities (Participate in activities), Activity restriction as an option to not worsen the pain (Activity restriction), Impaired enjoyment of activities together with family/friends (Enjoyment with family/friends), Changed relationships with relatives due to pain (Changed relationships), Pain impact on job satisfaction (Impact on job satisfaction), Pain impact on ability to do household chores (Ability to do household chores), The impact of pain on the ability to plan something in advance (Ability to plan), The impact of the pain on relationships with friends and acquaintances (Relation to friend/acquaintance).
Abbreviations: hEDS, hypermobility Ehlers Danlos syndrome; HSD, hypermobility spectrum disorders; IQR, interquartile range; *M*, mean; Md, median; MPI-S, multidimensional pain inventory Swedish version; *N*, numbers; RG, reference group; SD, standard deviation; %, percent.

TABLE 4 | Regression analysis for pain intensity (NPRS 0–10) in HSD/hEDS and RG.

	HSD/hEDS $N=1.185$					**RG $N=39.471$**				
	R^2	**B (CI for B)**	**p**	**t**	**df**	**R^2**	**B (CI for B)**	**p**	**t**	**df**
(Constant)	0.153	5.022 (4.180, 5.865)	< 0.001	11.693	3	0.151	5.423 (5.293, 5.554)	< 0.001	81.321	3
Number of pain locations		0.50 (0.38, 0.062)	< 0.001	7.926			0.45 (0.043, 0.047)	< 0.001	43.922	
Persistent pain		0.325 (−0.084, 0.735)	0.120	1.558			0.536 (0.471, 0.601)	< 0.001	16.234	
Recurring pain		−0.616 (−1.549, 0.228)	0.153	−0.129			−0.322 (−0.455, −0.188)	< 0.001	−4.707	

Abbreviations: *B*, unstandardized coefficients; df, degrees of freedom; hEDS, hypermobility Ehlers Danlos syndrome; HSD, hypermobility spectrum disorders; IQR, interquartile range; *M*, mean; Md, median; MPI-S, multidimensional pain inventory Swedish version; *N*, numbers; NPRS 0–10, numerical pain rating scale (pain intensity during the last week); Persistent pain and Recurring pain, question from health information "Is your pain periodically recurring or persistent yes/no?"; PRI 0–36, pain region index (number of pain locations); R^2, explained variance; RG, reference group; SD, standard deviation; *t*, *t*-test values indicate the predictors regression coefficient; %, percent.

were statistically significant differences, p-value <0.001, for pain characteristics (Table 2)—pain onset, pain duration, pain locations, and the pain intensity during the last week (NRPS, 0–10)—between the groups, where the HSD/hEDS group reported higher values, but the effect sizes (r) were less than 0.3 which indicated a small effect. These results are reported in more detail in Supporting Information (Table S2). In the HSD/hEDS group, 17.1% reported intermittent pain and 80.1% reported persistent pain; in the RG, 18.9% reported intermittent pain and 74.2% reported persistent pain. The HSD/hEDS group reported a pain onset ($M=13.5$, $\mathrm{SD}=10.9$) that was 4 years earlier than the RG ($M=9.5$, $\mathrm{SD}=9.8$ year), and the HSD/EDS group ($M=6.4$, $\mathrm{SD}=1.8$) reported lower pain intensity during the last week (NPRS) than the RG ($M=6.8$, $\mathrm{SD}=2.0$).

TABLE 5 | Regression analysis for interference in daily activities in HSD/hEDS and RG.

	HSD/hEDS $N = 1.211$					RG $N = 42.590$				
	R^2	B (CI for B)	p	t	df	R^2	B (CI for B)	p	t	df
(Constant)	0.278	1.67 (1.41, 1.93)	< 0.001	12.521	3	0.25	2.15 (2.11, 2.19)	< 0.001	99.638	3
Pain intensity during the last week (NPRS 0–10)		0.30 (0.26, 0.36)	< 0.001	15.759			0.29 (0.28, 0.3)	< 0.001	96.548	
Years since onset of pain		0.002 (−0.004, 0.008)	0.47	0.724			−0.001 (−0.002, 0.000)	0.043	−2.028	
Number of pain locations		0.03 (0.04)	< 0.001	6.291			0.02 (0.014, 0.017)	< 0.001	23.934	

Abbreviations: B, unstandardized coefficients; df, degrees of freedom; hEDS, hypermobility Ehlers Danlos syndrome; HSD, hypermobility spectrum disorders; N, numbers; NPRS 0–10, numerical pain rating scale (pain intensity during the last week); PRI 0–36, pain region index (number of pain locations); R^2, explained variance; RG, reference group; t, t-test values indicate the predictors regression coefficient.

Figure 1 illustrates the frequency distribution of pain locations for the HSD/hEDS and RG. The colour gradients in the figures reflect the two groups' ratings, in percentage, of the 36 pain locations in the PRI- index. The left image pair represents the HSD/hEDS group, and the right image pair represents the RG. Complete data are available in the Supporting Information (Table S2). The figures were drawn using a colour gradient from white to yellow (10%–50%) and from yellow to red (50%–90%). Both groups had similar pain patterns and reported pain in the lower back as the most painful followed by pain in the shoulders and pain in the hip/seat. Overall, the HSD/hEDS group ($M = 20.0$, $SD = 7.9$) reported more pain locations than the RG ($M = 14.8$, $SD = 8.8$).

The subscale MPI-S interference in daily activities (Table 3) measured pain interference with everyday activities. Statistically significant differences, p-value < 0.001, between HSD/hEDS and RG groups were found in 5 (participate in activities, activity restriction, enjoyment with family/friends, changes relationships and impact on job satisfaction) of the 11 items. Both groups showed similar scores, but the HSD/hEDS group generally appeared to have lower scores for pain interfering with daily activities, in relation to RG. Changed work ability since pain onset was reported as most affected by both the HSD/hEDS group (MD = 5.3, IQR = 4.1–6.0) and RG (MD = 5.4, IQR = 4.4–6.0). The lowest estimated change was reported regarding changed relationships, HSD/hEDS (MD = 3.4, IQR = 1.6–4.8), and RG reported (MD = 3.6, IQR = 1.9–5.0). More information can be found in the Supporting Information (Table S3).

Regression analyses were conducted to determine the relationship between various aspects of pain and the ability to perform daily activities.

The multivariable regression analyses (Table 4) indicated that the number of pain locations ($p < 0.001$) are significant for pain intensity during the last week for people with HSD/hEDS, but it is not crucial whether the pain is persistent ($p = 0.120$) or intermittent ($p = 0.153$). Statistical significance ($p < 0.001$) was indicated for the number of pain locations and intermittent or persistent pain for the RG-group.

The multivariable regression analyses (Table 5) indicate that both the pain intensity during the last week ($p < 0.001$) and the number of pain locations ($p < 0.001$) significantly explain the interference in daily activities for both the HSD/hEDS and RG groups. However, the onset of pain was not a significant predictor of interference in daily activities for the HSD/hEDS group ($p = 0.47$) or the RG group ($p = 0.043$). The variance inflation factor (VIF) values confirmed that there was no issue with multicollinearity in the regression analyses.

4 | Discussion

This study investigated aspects of pain and their impact on everyday life in people with HSD/hEDS. Despite similar patterns of activity interference due to pain (Table 3), the HSD/hEDS group reported less interference in activities enjoyed with family and friends and higher job satisfaction than the RG group (Table 3). People with HSD/hEDS developed pain at younger ages than people in the RG, with a mean age difference of 9 years. Various factors may contribute to the early development of pain in this group, including joint trauma during play or sports, repeated sprains, and subluxations, which can lead to early onset pain. Studies suggest that pain often starts locally as myofascial pain and may become more generalised, potentially evolving into nociplastic pain (Feldman et al. 2020; Syx et al. 2017; Tinkle 2020). The HSD/hEDS group also reported pain locations near joints such as shoulders, hips, and knees more frequently than the RG, likely due to joint instability and impaired proprioception, which are common in HSD/hEDS (Chopra et al. 2017; Fatoye et al. 2012; Tinkle 2020). This complex pain presentation, characterised by multiple pain types occurring simultaneously and changing over time, highlights the need to distinguish HSD/hEDS pain from other chronic pain conditions for optimal treatment (Chopra et al. 2017; Feldman et al. 2020; Ritelli et al. 2024). Our study found that the HSD/hEDS group reported significantly more pain locations and had a higher degree of persistent pain; yet surprisingly, they reported lower pain intensity compared to the RG. Previous studies have examined the relationships between pain intensity, the number of pain locations, and their impact on everyday life. Landmark et al. (2024) found that sleep disturbances and pain intensity are strongly associated with pain-related disability. Similarly, Gunnarsson et al. (2021) suggest that pain intensity significantly impairs the ability to engage in meaningful activities. In contrast, Mose et al. (2016) found that the number of pain locations appears to be more important in terms of sick leave than pain intensity. In this present study, we found that the number of pain locations could explain the pain intensity. However, no connection was found between the persistence or intermittency of pain in the HSD/hEDS group. In the RG, however, both the number of pain locations and the duration of the pain were found to influence pain intensity. This suggests that people in the RG may experience greater limitations in everyday life due to their pain intensity.

Our study showed that 2.8% of people in the SQRP had the diagnosis of HSD/hEDS, which is a higher incidence than that reported in a previous study, that also used data from the SQRP, where an incidence of 1.9% for people with HSD/hEDS was found between 1998 and 2016 (Molander et al. 2020). One explanation for this study's higher incidence compared to Molander's study may depend on updated diagnosis criteria in 2017 Furthermore, the available data is based on data from specialist pain clinics. A study from 2013 reports a prevalence of HSD/hEDS of approximately 3% in cross-sectional population survey (Mulvey et al. 2013), and another cohort study in Wales reports a prevalence of 1 in 500 för HSD/hEDS (Demmler et al. 2019). However, the prevalence in HSD/hEDS is uncertain, (Castori et al. 2017). There is a noticeable heterogeneity even within the diagnoses, which also complicates the conditions for making the correct diagnosis. Ritelli et al. (2024) reviewed 2017's new criteria (Malfait et al. 2017) and suggested a more broad-minded approach to the diagnoses, with the justification that people with HSD/hEDS would then have increased access to healthcare from more specialists and enabling an adequate assessment. This could promote the conditions of individuals with HSD/hEDS to receive a proper diagnosis, treatment, and rehabilitation, thereby enhancing their well-being and quality of life (Ritelli et al. 2024).

There is almost 10 years in mean difference in age between the HSD/hEDS group and the RG, with the HSD/hEDS group being younger. Perhaps, this is an explanation for the higher level of education and reported a lower impact of pain on work ability and job satisfaction in the HSD/hEDS group. Another explanation could be that the persons in the HSD/hEDS group have chosen professions that provide better conditions for dealing with work-related challenges. One could hypothesise that an onset of pain early in life could lead to adaptations at a younger age, to manage and navigate in everyday life. The range of daily activities for a 35-year-old can differ significantly from that of someone 10 years older. It may be easier for younger individuals to adapt, and thus, the HSD/hEDS group has had more opportunity to design and adapt their routines and habits based on their circumstances than older individuals, who possibly strive to return to their previous way of life. Adaptation is seen as a developmental process that continues throughout life. The adaptation process requires time, time to readjust, and create a new identity with new roles and habits (Dépelteau et al. 2021). Previous studies (De Baets et al. 2022, 2017) suggest that people with HSD/hEDS may adapt their daily activities over time to manage pain, which can support the findings in this study.

The results of our study can contribute to increased understanding and knowledge of the characteristics of pain and how activity limitations are expressed in people with HSD/hEDS. Still, further research is needed to understand how individuals with HSD/hEDS manage everyday activities and to determine if and how these activities contribute to or increase pain. This lack of understanding ultimately impacts our knowledge of how pain affects the quality of life for those with HSD/hEDS.

4.1 | Limitations

There are uncertainties in using data from quality registers that potentially constitute missing data. One uncertainty is how the register has handled data from both pain clinics and the participants who completed the questionnaires. We chose to use baseline data because we wanted to collect data from all patients available in SQRP. A study that reviewed 103 Swedish quality registers, including SQRP, found that 34 out of 34 clinics reported to SQRP with a completeness of 86% (Emilsson et al. 2015). Fischer et al. (2020) examined the coverage of reported data in the SQRP. The study found minor missing data for completed questionnaires from both patients, and the Pain clinics had also reported their data in a credible manner (Fischer et al. 2020), so we can assume that the data from the SQRP is as robust as possible.

Neither ICD-10-SE nor SQRP separate the 13 different EDS diagnoses but consider HSD and EDS as one group. Although the hypermobility variant of EDS is the most common variant (> 90%), it is possible that other EDS variants are included in our data. Additional uncertainties to consider are the significant prevalence of asymptomatic hypermobility, which is present in 10%–15% of the population. It is definitely possible that there are people who should have had an HSD/hEDS diagnosis who instead were placed in the RG. In 2017, new stricter diagnostic criteria for HSD/hEDS were introduced. It is unclear how adherence with the new guidelines is followed and whether it can affect the result in this study.

This study focuses on how pain affects everyday life, but other symptoms and co-morbidities can contribute to pain that impairs daily life activities, for example fatigue, dysautonomia, and mental illness (Hakim et al. 2021). The pain itself often leads to increased fatigue. Dysautonomia symptoms can further increase fatigue and pain with the risk of a poorer quality of life. Both overactivity and inactivity can contribute to increased pain and be part of pain management (Chopra et al. 2017; Simmonds 2022). There are also studies that show co-morbidity regarding mental or psychosocial health (Baeza-Velasco et al. 2018; Bulbena et al. 2017). It is possible that additional components influence the outcome of people with HSD/hEDS diagnoses, but the co-morbidity question is not addressed in this study.

In conclusion, people with HSD/hEDS experienced an earlier onset of pain, reported more pain locations, and had more persistent pain compared to those in the RG. Pain duration and the number of pain locations significantly impacted their daily activities, although pain intensity was less decisive. Further research is essential to better understand how pain affects valued activities and the daily lives of people with HSD/hEDS and to develop targeted interventions for improving their quality of life.

Author Contributions

All authors have participated in the design of the study, discussed the results, and commented on the manuscript.

Conflicts of Interest

The authors declare no conflicts of interest.

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Supporting Information

Additional supporting information can be found online in the Supporting Information section.