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### Mind the gaps

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## Original article

# Mind the gaps: therapists' experiences of managing symptomatic hypermobility in Scotland

Dervil M. Dockrell \*, Kathryn M. Berg<sup>†</sup>\* and Stuart H. Ralston

## Abstract

**Objectives** The aim was to ascertain occupational therapist (OT) and physiotherapist (PT) experiences of managing hypermobility spectrum disorders (HSDs) and hypermobile Ehlers-Danlos syndrome (hEDS) patients, specifically the training and confidence levels of therapists, use of evidence-based practice, accessibility of pain management and clinical psychology services, and perceived usefulness of a specialist centre in Scotland.

**Methods** A mixed-method survey was distributed to Rheumatology OTs/PTs in Scotland. It included multiple choice and open text questions, which were analysed to reflect therapists' experiences and perception of service need.

**Results** We found that therapists in Scotland do have expertise in the management of HSD/hEDS patients; however, this expertise tends to be concentrated in secondary care, which makes it difficult for patients who are managed in primary care to access. The majority of respondents reported lack of access to external training (80%). There was difficulty in referral to pain management services (55%) and clinical psychology (28%) among adult therapists. Paediatric services provided considerably better access to these disciplines. Of note, the majority of respondents were in favour of a specialist centre in Scotland for the training and education of therapists (94.7%) and the diagnosis and management of complex HSD/hEDS patients (73.7%).

**Conclusion** More research is needed urgently to evaluate the effectiveness of therapy interventions to underpin a national guideline in order that we can improve outcomes for HSD/hEDS patients. A specialist centre with expert and engaged clinicians would be a valuable asset in coordinating patient-focused research and conducting good-quality clinical trials.

**Key words:** occupational therapist, physiotherapist, hypermobile spectrum disorder, hypermobile Ehlers-Danlos syndrome, Scotland

## Key messages

- Despite therapy expertise, there is still unmet need and discrepancy in service provision for hypermobility spectrum disorder/hypermobile Ehlers-Danlos syndrome patients in Scotland.
- A specialist centre for this patient group in Scotland would be valuable.
- More research is required to underpin national therapy guidelines for treatment of adult hypermobility spectrum disorder/hypermobile Ehlers-Danlos syndrome patients.

Centre for Genomic and Experimental Medicine, MRC Institute of Genetics and Cancer, University of Edinburgh, Edinburgh, UK

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\*Dervil M. Dockrell and Kathryn M. Berg contributed equally to this manuscript.

Correspondence to: Dervil M. Dockrell, N3.16, Centre for Genomic and Experimental Medicine, MRC Institute of Genetics and Cancer, University of Edinburgh, Edinburgh, UK. E-mail: dervil.dockrell@ed.ac.uk.

## Introduction

Joint hypermobility is defined as the ability to move joints beyond the normal range of movement [1]. For some individuals it can cause symptoms of pain, swelling, stiffness and subluxation, which can adversely affect quality of life [2]. The Ehlers-Danlos syndromes (EDS) are a group of heritable connective tissue

disorders caused by mutations in genes that encode collagen and other molecules involved in maintenance of soft tissues [3]. The 2017 International Classification of EDS refined the criteria for the 13 types of EDS, of which hypermobile EDS (hEDS) is a subtype. Unlike the other forms, hEDS is thought to be polygenic in nature and cannot be diagnosed by molecular genetic testing. It is characterized by joint hypermobility, skin flexibility and tissue fragility [4]. Where criteria are not met for hEDS or any other connective tissue disorder, a diagnosis of hypermobility spectrum disorder (HSD) may be given.

HSD and hEDS can significantly impair health and wellbeing and present a challenge for health professionals to manage [5–8]. Recent work suggests that these diagnoses can affect many systems [9], not only the musculoskeletal system [10–15]. The presence of non-musculoskeletal symptoms is not included in the diagnostic criteria of hEDS, but these conditions can be more limiting than the musculoskeletal symptoms themselves [16].

It has been acknowledged that the availability and quality of services for patients with HSD/hEDS in the National Health Service (NHS) varies widely [17]. The time from onset of symptoms to diagnosis and treatment can be lengthy, which can have a negative impact on patient outcomes [12]. In 2017, the Scottish Government committed to reviewing services for EDS patients in response to patient feedback that indicated an apparent lack of coordinated care and access to treatment [18].

EDS Support UK does not verify the diagnoses of their members but has recorded 466 patients in Scotland with hEDS and 60 with HSD (Julier, K., Ehlers-Danlos Support UK, 2020 written communication, 1 December). It is thought that the true prevalence of HSD/hEDS is unknown because it is widely underdiagnosed despite symptoms presenting commonly in rheumatology clinics [19]. In 2017, the charity conducted a survey of its patient members in Scotland. The survey recorded 19 years being the average length of time between symptom presentation and diagnosis in Scotland. Eighty-seven per cent of respondents felt that their condition was not well supported by the NHS. Eighty-three per cent reported that they did not have access to a medical professional knowledgeable about their condition [20].

A 2020 report on behalf of EDS Support UK discusses the experience of young people with HSD/EDS in Scotland. The report states that young patients in Scotland perceive the attitudes of health-care professionals to be negative. This is particularly prevalent in young females, who report that their symptoms are not believed. The diagnostic journey for EDS/HSD patients is problematic, and the report revealed an inequity of service access in Scotland, with most patients surveyed reporting that they had to travel to London to receive a diagnosis. The report advocates for better education and training of health professionals in Scotland and the need for better service access geographically [21].

Although there are several reports related to the patient experience of the management and diagnosis of

HSD/hEDS in Scotland, the viewpoints of therapists are underrepresented in the literature. The primary aim of this study was to capture the perceptions and experiences of occupational therapists (OTs) and physiotherapists (PTs) who manage patients with HSD and hEDS in Scotland. The secondary aim was to ascertain whether there is a need for a specialist centre in Scotland.

## Methods

A cross-sectional mixed-methods questionnaire was designed (see [Supplementary Data S1](#), available at *Rheumatology Advances in Practice* online). A mixture of Likert scales, checkboxes and descriptive questions were used. It was determined that a purposeful sampling method was most appropriate for this study.

The questionnaire was constructed to gather information on the facilities and resources available to therapists working with hypermobility, the extent to which they were confident in assessing those patients, the techniques and outcomes they used and the clinical characteristics of their caseloads. The survey questions reflected interventions recommended in the literature for treating rheumatology patients [22–25].

The survey respondents were identified as members of registered professional bodies currently managing patients with symptomatic hypermobility in Scotland. OTs and PTs who were known to be members of registered professional bodies such as the British Society for Rheumatology (BSR), the Scottish Paediatric and Adolescent Rheumatology Network, and the Royal College of Occupational Therapists Rheumatology Clinical Forum were invited to complete the survey, because these bodies were determined to be most likely to represent allied health professionals treating these patients.

A total of 20 responses representing 11 of the 14 Scottish health boards were received. Eighty per cent of the respondents worked in hospitals, representing a secondary care service. Fifty-five per cent of the respondents treated adults and young people [16–25]; the other 45% treated paediatric patients  $\leq 19$  years of age.

Data were coded in Excel. The following themes were identified: referrals and appointments; clinical practice; therapist proficiency; other service access; and service needs. Statistical significance was determined using Fisher's exact test in Minitab 17 software to compare categorical values. Ethical approval was not required for this study as determined by the UK policy framework for Health and Social Care Research.

## Results

### Referrals and appointments

The most common referral sources reported by respondents were through rheumatology teams (95%). Sixty per cent of therapists also reported referrals from

TABLE 1 Treatment interventions used by therapists

Treatment intervention	Occupational therapist (n = 13)		Physiotherapist (n = 7)		All therapists (n = 20)	
	n	Percentage	n	Percentage	n	Percentage
Energy conservation principles	13	100	7	100	20	100
Self-management	12	92	7	100	19	95
Joint protection techniques	12	92	3	43	15	75
Sleep hygiene	10	77	5	71	15	75
Home exercise programme	5	39	7	100	12	60
Hand therapy	8	62	2	29	10	50
Motivational interviewing	6	46	4	57	10	50
Relaxation skills	6	46	4	57	10	50
Cognitive behavioural approach	4	31	3	43	7	35
Other*	2	15	2	29	4	20

\*Other includes hydrotherapy, mindfulness, yoga, tai chi and Pilates.

general practitioners (GPs). Other referral sources reported were orthopaedics (25%), cardiology (10%), gastroenterology (5%), genetics (5%) and schools (10%).

Most of the respondents (65%) reported that their referrals did not use the 2017 terminology. Other terminology used included the terms JHS, BJHS, EDS type III and EDS hypermobility type.

Therapists were asked how many HSD/hEDS patients they treated in a usual calendar month. Most (60%) therapists saw fewer than five HSD/hEDS patients per month. Twenty-five per cent of therapists had a caseload of between 5 and 10 patients, and 15% saw >10 patients per month. Twenty-five per cent of therapists reported that they see other types of EDS patients.

Two respondents used free text to highlight the lack of specific hypermobility referrals in their caseloads:

"[Hypermobility] would only be as a secondary diagnosis to an inflammatory arthritis in most cases, we do not take referrals purely for the diagnosis of hypermobility. We would take a referral for EDS and more complicated cases but these are rare." Respondent number 6.

"Hypermobility is often not the primary diagnosis on the referral." Respondent number 5.

Thirty-five per cent of respondents reported that more than half of their caseload was represented by patients with complex needs. The complex needs reported can be found in the questionnaire in the [Supplementary Material](#), available at *Rheumatology Advances in Practice* online.

All respondents reported that they were able to offer more than one appointment for patients under their care. Six therapists (30%) reported that >50% of their patient caseload required four or more sessions.

### Clinical practice

A copy of the questionnaire used and a list of the assessment areas surveyed can be found in the [supplementary material](#) (see [Supplementary Table S1](#), available at *Rheumatology Advances in Practice* online).

Common areas of assessment (represented by  $\geq 85\%$  of all therapists) were: work and education (95%); exercise and hobbies (95%); and fatigue (95%).

Any differences between OTs and PTs were determined to be discipline related; for example, OTs were more likely to assess functional activities, such as personal activities of daily living, domestic activities of daily living, handwriting and computer use, whereas PTs were more likely to assess areas such as muscle length, muscle strength, gait and balance.

[Table 1](#) summarizes the types and popularity of treatment interventions used by therapists when treating HSD/hEDS patients. As with the assessments, differences between OT and PT treatment interventions were determined to be discipline specific; hence, statistical analysis was considered to be redundant.

One respondent indicated that a lengthy diagnosis may act as a barrier to effective treatment of HSD/hEDS patients in their caseload:

"My feeling is that referral onto me is often too late as individuals have already well-established bad habits". Respondent number 1.

Fifty-seven per cent of PTs and 62% of OTs reported that they used outcome measures to assess the effectiveness of treatment in patients (see [Supplementary Table S2](#), available at *Rheumatology Advances in Practice* online, for a list of the types of outcome measures reported).

### Therapist proficiency

When asked what training they had received to assess and treat HSD and hEDS patients, six respondents (30%) had received external training (see [Supplementary Table S3](#), available at *Rheumatology Advances in Practice* online), four (20%) therapists had visited other centres, five (25%) reported that they had received in-house training, and 95% reported that they had undertaken self-directed training. Of those who worked with young people and paediatric patients, 50% reported that they were aware of the British Society for Paediatric

**TABLE 2** Subjective confidence levels of therapists in the management of HSD/hEDS patients

Confidence level	OT		PT		Overall		P-value
	n	Percentage	n	Percentage	n	Percentage	
Very confident	0	0	2	29	2	10	<i>P</i> = 0.110
Moderately confident	9	69	5	71	14	70	<i>P</i> = 1.000
Not confident	4	31	0	0	4	20	<i>P</i> = 0.248

Significance was determined as  $P < 0.05$ . No significance was found when calculated using Fisher's exact test. OT: occupational therapist; PT: physiotherapist.

and Adolescent Rheumatology (BSPAR, now integrated into BSR) guidelines for the assessment and treatment of symptomatic hypermobility.

There were no statistically significant differences between the confidence levels of OTs and PTs in dealing with HSD/hEDS, despite the fact that 31% of OTs reported that they were not confident in the management of patients with HSD/hEDS. Overall, 80% of therapists reported that they were confident in the assessment and treatment of HSD/hEDS (Table 2).

#### Respondent-reported service access

Few therapists (55%) managing adults had access to a pain management programme compared with 89% of therapists managing paediatric patients, but the difference was not statistically significant when calculated using Fisher's exact test ( $P = 0.157$ ).

Fewer therapists managing adults reported access to a clinical psychologist (28%) compared with 89% of paediatric therapists, and this was highly significant when calculated using Fisher's exact test ( $P = 0.009$ ; see Fig. 1).

#### Need for a specialist centre

The majority of respondents were in favour of a specialist centre for the management and treatment of patients and for the training and education of therapists (see Figs 2 and 3). However, one therapist noted that if HSD/hEDS patients were discharged back to a local centre for the implementation of their treatment plan, they would be unable to absorb the extra workload. This suggests that there is a need for further investment in recruitment and training of additional specialist therapists:

"I do not have the capacity to see increased patient numbers, especially if there is an expectation on the part of all patients with EDS going through such a centre." Respondent number 1.

## Discussion

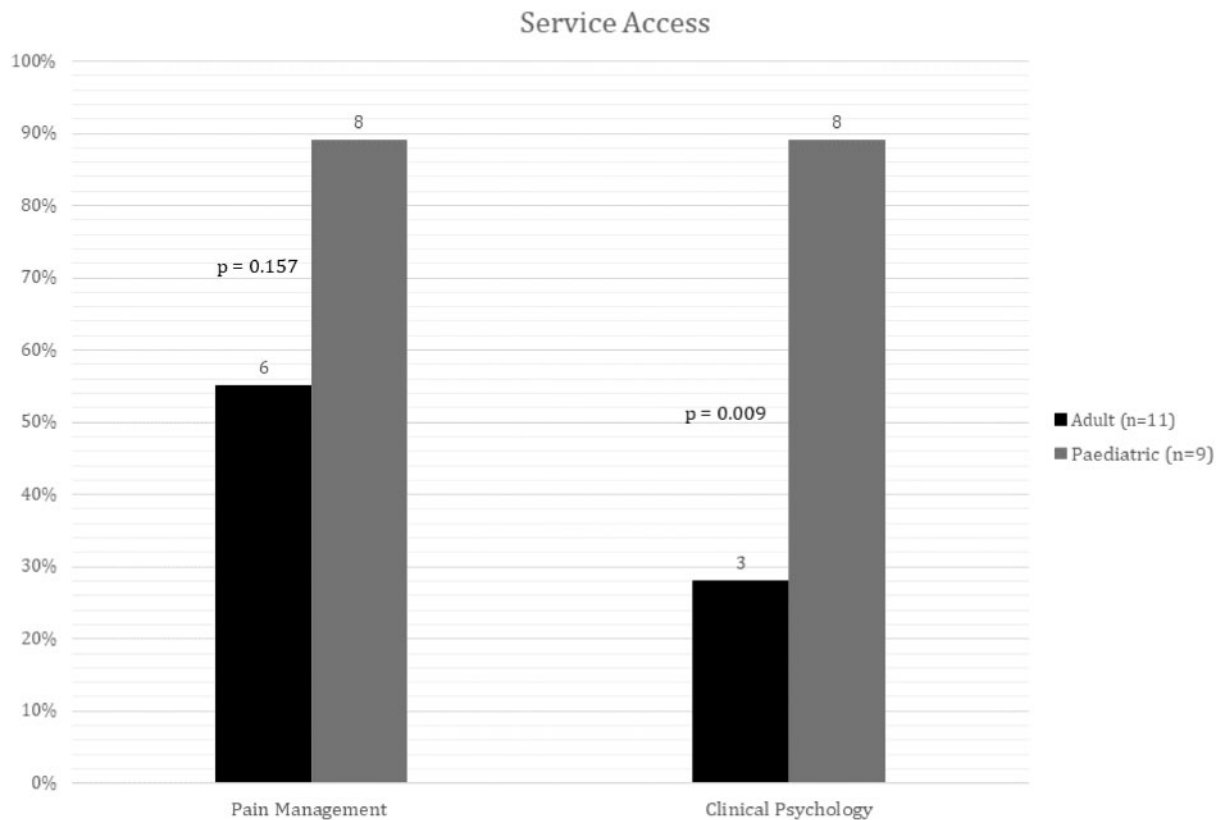
The purpose of this study was to collate the experiences of Scottish rheumatology therapists in the management of patients with HSD and hEDS and to ascertain whether there is a need for a specialist centre. In the

absence of specific guidelines for OTs on management of adult patients with HSD and hEDS, the literature recommends OT as part of an MDT approach to managing patients [1]. The biopsychosocial model of care, which is the core approach for OTs, is advocated by Baeza-Velasco *et al.* [26] for patients with hypermobility. Hammond [27] reports self-management programmes, joint protection education, fatigue and mood management, activity and role planning, goal setting and counselling as OT interventions for chronic musculoskeletal conditions.

All OTs used the recommended treatments, but some of these were used by as few as one-third of therapists. PT respondents reported similar adherence to the evidence base for treatment strategies, with the emphasis on active vs passive treatment. More than half (57%) did not report using specific joint protection techniques as advocated in the literature [28, 29].

Current therapy management is largely the same for HSD and hEDS, although there is a paucity of quality literature on the clinical and economic effectiveness of interventions. Russek *et al.* [25] outlined the diagnosis and management of patients with HSD and hEDS for PTs. They explored the terminology of the diagnoses and complexity of the condition. Evaluation and treatment strategies for patients are recommended, with an emphasis that therapists who are knowledgeable about the conditions should provide such treatment. Owing to the limited research on the best therapeutic interventions, therapists should rely on their clinical experience and patient partiality.

More than one-third of the therapists did not use quantitative outcome measures to evaluate patient treatment, and only one respondent indicated that they used a specific hypermobility outcome measure for hypermobility. This is reflective of the lack of evidence in the literature regarding effectiveness of interventions [25, 28]. Where outcome measures are used (see Supplementary Material, available at *Rheumatology Advances in Practice* online), they are consistent with those reported in the survey by Palmer *et al.* [30] of PTs in 2015, indicating that practice is evidence based. The majority of the therapists reported that they were moderately confident in the assessment and treatment of patients with HSD/hEDS, but only 20% had previously received any specialist training. Nearly all (94.7%) respondents were

**Fig. 1** Respondent-reported access to pain management and clinical psychology services

in favour of a specialist centre for education and training in the management of HSD/hEDS.

Few of the therapists who were dealing with adult patients had access to a clinical psychologist. This is important because anxiety and depression are highly prevalent symptoms in this patient group, and it is likely that they are not being addressed appropriately [24]. This is in contrast to the paediatric population, in which nearly 90% of therapists reported access to this service. It has been demonstrated that psychological distress is significantly higher in patients with hypermobility than in those without [31]. Although pain has a moderate impact on disability, fatigue, anxiety and depression have more of an impact on health in this patient group [32]. A study by Bennett *et al.* [33] concluded that there was a need to heighten awareness of hypermobility in the health-care professional to improve outcomes for patients.

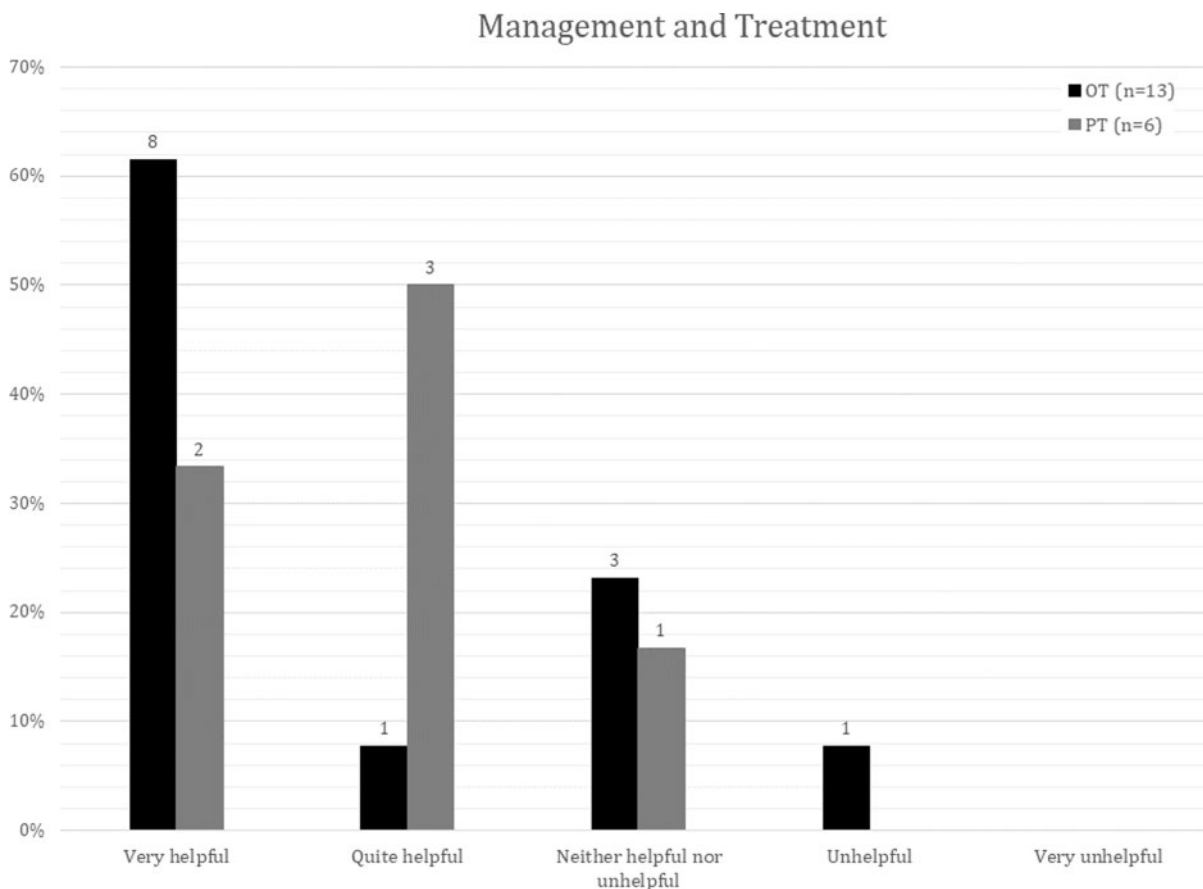
Clinical psychology is a valuable resource in the treatment of HSD/hEDS. Patients have a higher likelihood of anxiety, depression and panic disorders [34] and report feelings of guilt and low self-esteem, often resulting from a poor perception of the condition among the health-care community [35]. Patients often report a lengthy wait between the onset of symptoms and the diagnosis of HSD/hEDS and may see a number of

different health professionals. This can exacerbate feelings of anxiety and depression. A 2016 study by Schepers *et al.* [32] suggested that emotional health in this patient group influenced their wellbeing more than physical discomfort.

Only 55% of therapists treating adults reported access to a pain management programme, compared with 89% of those managing paediatric patients. Pain management services provide settings in which patients are offered a rehabilitative psychological approach to pain self-management. The programme can be individual or group based. Rahman *et al.* [36] developed a condition-specific pain management programme for hypermobility patients. The results of several outcome measures suggested that improvement was substantial and continuous. It is our view that such a service would be fundamental in the management of HSD/hEDS patients.

Despite the clarifications made in the 2017 EDS International Classification regarding the nomenclature of HSD/hEDS, the majority of our respondents (65%) reported that the referrals they received from services used the older terminologies. The authors suggest that this continued use of previous nosology could demonstrate a lack of awareness of the current classification criteria and recent literature among the various medical

**Fig. 2** Percentage of therapists who answered 'Yes' to the question, 'How useful would a specialist centre for the management and treatment of patients be?'



specialities who referred patients to therapy. However, it should be noted that if hypermobility was not newly diagnosed at the time of referral, then historical terminology might be used.

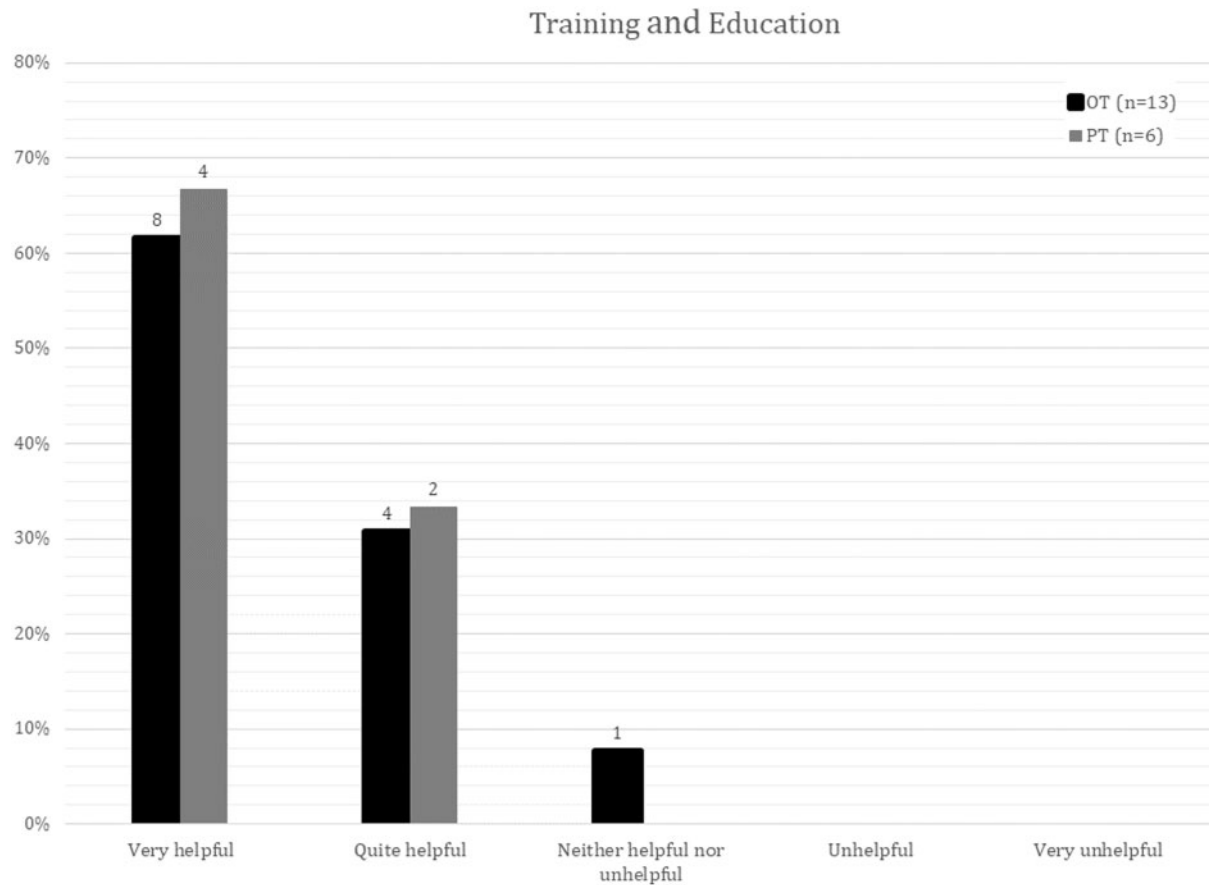
These results suggest that therapists do assess and treat HSD and hEDS patients according to the available evidence base. However, it is clear that more training and education are required for therapists to assess and treat patients consistently across Scotland. The majority (94.7%) of therapists were in favour of a specialist centre to provide this service. This is bolstered by the 2020 EDS Support UK report, which revealed an inequity of service access in Scotland [21].

The BSR issued a statement in March 2020 concluding that there was insufficient evidence for them to develop a guideline for the management of hypermobility at present [37, 38]. The Royal College of General Practitioners (RCGP) toolkit was developed in conjunction with EDS UK in 2018 [39], but raised safeguarding issues for children with the diagnosis [37, 40]. This toolkit was to be removed from the RCGP website but will remain for the present (Ehlers-Danlos Support UK, 2021, written communication updated 1 February 2021).

The Hypermobility Syndromes Association (HMSA) Kent Model provides training resources for the health professional [41]. The EDS Society runs Project ECHO, with training courses for professionals and allied health professionals [42, 43]. The Scottish Paediatric Adolescent Rheumatology Network (SPARN) has a recommended pathway of care for children with joint hypermobility [44]. In 2019, the allied health professional members of BSPAR reviewed their guidelines for management of children and young adults with symptomatic hypermobility [45]. These guidelines have been criticized by members of EDS patient support groups in the USA and the UK for a number of reasons, including the assertion that difficulties occur mainly when the body is weak, the guidance that gastrointestinal and urinary involvement is rare, and the fact that patient groups were not involved in the development of the recommendations [46].

Despite the availability of information on the management of HSD/hEDS, the reality is that accessing appropriate health-care support is very difficult for patients in Scotland [21]. The recommended pathway for patients with symptomatic hypermobility is diagnosis by GP

**Fig. 3** Percentage of therapists who answered 'Yes' to the question, 'How useful would a specialist centre for the training and education of therapists be?'



followed by self-care information, physiotherapy and exercise, occupational therapy, podiatry, pain killers and pain management. Other systemic problems may also be diagnosed by the GP or referred to a hospital speciality [47].

In the absence of a specialist centre, National Services Division Scotland (NSD) commissions a highly specialist diagnostic service for complex EDS patients through their block agreement with NHS England. The NHS England-commissioned service comes into play for very difficult cases. Although this service offers management advice, there is no access to specialist PTs or OTs. This diagnostic service is highly specialized, with narrowly defined acceptance criteria, which most hEDS/HSD patients in Scotland would not meet, limiting patient access to the service. Access is also limited by a reluctance to refer by clinicians, who may not know that the service exists or may not feel that the condition warrants specialist diagnosis.

A few cases are referred each year to specialist residential programmes offered by a number of trusts in

England. These programmes offer access to specialist PTs and OTs with extensive experience in treating this cohort. Given that these services are not commissioned by NHS England, NSD does not normally fund referrals, although funding has been authorized in the past for some patients. The number of Scottish patients accessing these programmes may be considerably higher, because individual health boards may have opted to fund referrals for some of their patients without notifying NSD. In addition, patient organizations have highlighted that many patients have accessed these programmes through private insurance or chosen to self-fund (Roexe, A., National Services Division Scotland, 2021, written communication, 22 January).

It is clear from the results of the present study and from recent work published by EDS UK that an equivalent centre in Scotland would be welcomed by the majority of therapists and patients. The current pathways of care present challenges for HSD/hEDS patients, and there is a poor representation of relevant and necessary professions managing this patient group. These patients



are substantial users of secondary care services in NHS Scotland, and yet their needs remain unmet. It is our opinion that an investment in the diagnosis and management of HSD/hEDS is long overdue in Scotland and must be addressed imminently.

## Conclusion

The study has some limitations. The sample size was small and limited to musculoskeletal therapists working with hypermobility patients in Scotland. Surveying a larger sample of therapists would be of interest, particularly those working outside rheumatology.

We conclude that there is OT and PT expertise in management of HSD and hEDS patients in Scotland. A specialist centre would be welcomed by a majority of therapists. Important services, such as pain management and clinical psychology, are difficult to access.

Current specialist PT and OT services are mainly accessed through or concentrated in secondary care. The authors recommend an investment in primary care capacity for this patient group, particularly in the availability of specialist OT and PT services, which are poorly represented in primary care. A combined OT/PT approach would provide more holistic patient management. It is our opinion that clinicians in a specialist centre would be well placed to provide training and education to bolster the expertise of a primary care workforce.

For complex cases, clinicians in a specialist centre would also be able to develop a patient management plan to be maintained in primary care. This would lessen the burden on secondary care services and provide local access for patients. The availability of pain management and clinical psychology services is inequitable between adult and paediatric services. All patients should have access to these services, preferably in the community but at the very least through secondary care or a specialist centre.

More research is needed urgently to evaluate the effectiveness of interventions to underpin a national guideline in order that we can improve outcomes for these patients. A specialist centre with expert and engaged clinicians would be a valuable asset in coordinating patient-focused research and conducting good-quality clinical trials.

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## Data availability statement

The data underlying this article will be shared on reasonable request to the corresponding author.

## Supplementary data

Supplementary data are available at *Rheumatology Advances in Practice* online.

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