Exercise Beliefs and Behaviours of Individuals with Joint Hypermobility Syndrome/ Ehlers Danlos Syndrome-Hypermobility Type

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Abstract

Purpose: To explore exercise beliefs and behaviours of individuals with Joint Hypermobility Syndrome/ Ehlers Danlos Syndrome – Hypermobility Type and to explore patient experiences of physiotherapy.

Methods: A cross sectional questionnaire survey design was used to collect quantitative and qualitative data from adult members of the Hypermobility Syndromes Association and Ehlers Danlos Syndrome Support UK. Descriptive and inferential statistics were used to analyse the data. Qualitative data was analysed thematically.

Results: 946 questionnaires were returned and analysed. Participants who received exercise advice from a physiotherapist were 75.3% more likely to report high volumes of weekly exercise (Odds Ratio = 1.753% Confidence Interval = 1.30-2.36, $p < 0.001$) than those with no advice. Participants who believed that exercise is important for long term management were 2.76 time (276%) more likely to report a high volume of weekly exercise compared to the participants who did not hold this belief (Odds Ratio = 2.76, 95% Confidence Interval = 1.38-5.50, $p = 0.004$). Three themes emerged regarding experience of physiotherapy; physiotherapist as a partner, communication – knowledge, experience and safety.

Conclusion: Pain, fatigue and fear are common barriers to exercise. Advice from a physiotherapist and beliefs about the benefits of exercise influenced the reported exercise behaviours of individuals with Ehlers Danlos Syndrome – Hypermobility Type in this survey.

Keywords: Joint Hypermobility Syndrome, Ehlers Danlos Syndrome – Hypermobility Type, Exercise Beliefs, Exercise Behaviour, Physiotherapy, Physiotherapy

Introduction

Joint Hypermobility Syndrome (JHS) and Ehlers Danlos Syndrome – Hypermobility Type (EDS-HT) have been described as two heritable connective tissue disorders principally characterized by generalized joint hypermobility, complications of joint instability, skin laxity and fragility and chronic musculoskeletal pain (1). Furthermore, fatigue, muscle weakness and muscle cramps are commonly associated features which contribute to reduced quality of life and disability (2–4). Originally considered as two distinct conditions, many clinicians and researchers have interpreted JHS and EDS-HT as an expression of the same disorder (5). In this paper the term JHS/EDS-HT is used to describe these overlapping conditions (5).

Although the epidemiology of JHS/EDS-HT has not been thoroughly explored, the prevalence in the general population has been estimated between 0.75% and 2% (6) with women being much more frequently effected than men (1-5). Reports from clinical settings suggest a much higher prevalence of JHS/EDS-HT of between 30% and 60% in adult populations (7–10).

The clinical picture attributable to JHS/EDS-HT is still emerging. At the time when the Brighton (11) and Villefranche criteria (12) were established, both disorders were considered as mutually exclusive musculoskeletal disorders with cutaneous involvement. Significant disability has been identified in individuals with JHS/EDS-HT including walking, running, stair climbing, sport participation and personal hygiene (13–15). A recent meta-analysis showed that pain, fatigue and psychological distress had a significant impact on disability (15). Furthermore more complex multisystem involvement has been identified including autonomic and cardiovascular (16–18), respiratory (19), gastrointestinal (20,21),

genitourinary (22) and visual systems (23) although these relationships have not been proven to be causal. Clinical research is in its infancy and consequently optimal management JHS/EDS-HT is not yet defined.

Physiotherapy is considered a cornerstone of treatment however reports of physiotherapy management are not always favorable. With patients reporting that physiotherapy has exacerbated symptoms and have focused on one single joint rather than treating them holistically (24,25). Education, reassurance, closed chain strengthening and core stability exercises are recommended by experts and used frequently by physiotherapists (26). However while muscle strengthening and proprioceptive exercises have shown promising results for reducing pain and increasing strength in the knee (27,28) high quality intervention studies addressing mental health, widespread pain and fatigue are lacking (15,29,30).

Therefore, while exercise is a common component of treatment and appears to provide some benefits, there is has been no explorations of the patient experience of exercise interventions. A greater understanding of individual preferences, perceptions of exercise and experiences of physiotherapy may help to optimise the treatment approach and help to inform future research interventions.

The aim of this study was to explore the beliefs, attitudes and behaviours of individuals suffering with JHS/EDS-HT towards exercise and to explore their experiences of physiotherapy.

Methods

A cross sectional questionnaire survey design incorporating open and closed questions was undertaken.

Participants

Participants were adults aged 18 years or over with a reported diagnosis of JHS and or EDS-HT. Recruitment was via the Hypermobility Syndromes Association (HMSA) and Ehlers Danlos Syndrome Support UK (EDS UK) patient support groups. An online questionnaire with an invitation to participate and completion instructions were posted on the organisations' Website and Facebook pages. To ensure all members without internet access had the opportunity to participate, questionnaires were also posted to members with stamped addressed envelopes. Participants were included is they were 18 years or older and had received a diagnosis of JHS or EDS-HT. They were excluded if they reported diagnoses of other hypermobility syndromes such as Sticklers Syndrome, Marfan Syndrome, Osteogenesis Imperfecta, or other forms of EDS. The research was approved by the School of Health and Emergency Professions Ethics Committee, University of Hertfordshire.

Questionnaire development

A 21 item self-administered questionnaire comprised of both open and closed responses was developed. The questionnaire was designed after careful consideration of a questionnaire used to explore similar patient (Fibromyalgia) group perceptions of exercise (31). The specific aims of the current survey were considered and key features of the previous tools were selected and adapted to address those aims. A wide range of textbooks, scholarly articles, reviews and original research related JHS/EDS-HT were also consulted to inform the selection of specific questionnaire items. A draft questionnaire was developed and distributed for critique by three specialist physiotherapists each with more than 15 years of experience treating individuals with JHS/EDS-HT, three rheumatology consultants with

more than 20 years of treating individuals with JHS/EDS-HT and two patient representatives to further ensure face validity. The questionnaire was then modified in response to these suggestions. The final version (See supplementary material) of the survey addressed the following domains:

- Demographics and clinical characteristics
- Exercise beliefs
- Exercise behaviours and barriers
- Experiences of physiotherapy

Data management and analysis

Data was coded and transferred to Microsoft Excel and scrutinised by 2 members of the researcher team and later transferred to IBM Statistical Package for the Social Sciences (SPSS) (IBM Corp. Released 2015. IBM SPSS Statistics for Windows, Version 23, Armonk, NY: IBM Corp.). Agreement regarding categories for self-reported comorbidity data was obtained from co-author specialist physicians. Analyses were conducted on demographic and aggregated belief data to determine the most important factors which influenced the volume of exercise that a person reported undertaking each week. There were five categories of reported exercise volume (no exercise, < 30 minutes per week, 30-60 minutes per week. 1 hour – 2.5 hours per week, > 2.5 hours per week). These categories were selected in order obtain an overview of volume of exercise undertaken by the patient group. To avoid ambiguity and confusion with regard to terminology for the patients the term ‘exercise’ was used consistently in the questions, where technically the term ‘physical activity’ should have been used in accordance with the definition provided by Casperon and colleagues in 1985 and subsequent work in this field(32). Pearson Chi-Square test was used to test the association by determining the frequency and percentage for each of the different of activities with each of the independent categorical variables. Spearman’s rank correlation

test was used to test the correlation between the participant belief variables against each of the volume of exercise categories. An ordinal regression analysis was used to determine the probability of the participants' volume of exercise using the odds ratio. Selected factors influencing exercise behaviour and commonly reported symptom variables were subsequently included in the multiple logistic regression analysis. All significant demographic variables (i.e. employment and exercise advice) and all the statistically significant Spearman's rank correlated variables with a correlation coefficient greater than 0.15 (i.e. poor balance, belief in exercise for long term management, belief in exercise for control pain, belief in exercise for wellbeing and belief in exercise for mental functioning) were chosen as predictors for the ordinal regression model. The logit link function was used in the ordinal regression modelling. Thus, coefficients in a logit regression model are interpreted in terms of log-odds. The model fitting information implies that the relationship between volume of exercise and the independent variables of the model provides better predictions than guessing ($\chi^2 = 200$, $df = 17$, $p < 0.001$) (33). Although the pseudo R-square value (Nagelkerke $R^2 = 0.201$) was low, it never the less did indicate that a proportion of the outcome variable (exercise volume) was accounted for by the predictive variables of the model. For the goodness-of-fit test of the model, the p-values of the Pearson's and Deviance's chi-square tests were 0.150 and 1.000 respectively, which indicated there were no issue with the data fitting the model (34).

The open question responses regarding the participants' experience of physiotherapy was coded using a line by line approach. The codes where then grouped into categories. The codes and categories were then reviewed by a second reviewer and themes were then agreed (35).

Results

A total of 948 completed questionnaires were received (n = 432 paper responses and n = 516 online responses). Two duplicate questionnaires were removed, giving a total of 946 responses. Twelve incomplete questionnaires were included in the data analysis. These incomplete questionnaires were included, because 15 or more questions were answered and the data could be analysed was meaningfully.

Demographic and clinical characteristics

The majority of participants were female, 95.8% (n = 906), white 96.4% (n = 912) and 72.1% were aged between 18 and 40 years of age (67%). Forty-two percent (n = 399) were single, divorced/ separated or widowed. The majority, 81.3% (n = 763) were educated at University level. At the time of the survey 45.6% (n = 430) of participants were in full or part time employment, while 24.5 % (n = 231) were not working due to ill health. Fifty percent (n = 473) had experienced symptoms for 20 years or more. See Table 1. The spine (lumbar, thoracic and cervical) was the most problematic and most commonly reported region 60% (n = 566) with the knee 58 % (n = 545), hip 52% (n = 488), shoulder 52 % (n = 456) and wrist/hands 51% n = 444) also commonly cited. See Figure 1 for further detail. Comorbidities within this population were frequently reported, with 80% (n = 753) reporting that they had been diagnosed with other medical conditions. Mental health 43.4% (n = 411), musculoskeletal 42.8% (n = 405), cardiorespiratory including cardiac dysautonomia 41% (n = 385) gastrointestinal 27% (n = 251) conditions were the most commonly reported categories. Table 2 provides a summary of the most commonly reported co existing medical conditions.

Factors influencing exercise behaviour

The values for the Pearson chi-square (χ^2) test results presented in Table 3, show that the reported volume of weekly exercise was associated with having received exercise advice

from a physiotherapist ($\chi^2 = 26.85$, $df = 4$, $p < 0.001$). Also the participants' employment status was associated with the volume of exercise reported by participants ($\chi^2 = 14.45$, $df = 8$, $p < 0.001$). Moreover, the value of the Spearman's rank correlation test showed a significant but low correlation between the level of education and the reported volume of exercise ($r = -0.08$, $p = 0.014$).

The majority of participants "agreed" with the statements that exercise is important for fitness 90% (n = 850), wellbeing 78% (n = 741) and long term management of JHS/EDS-HT 77% (n = 701). Less than one half of participants 42% (n=339) agreed that exercise was important for pain management, while 58% (n = 607) were ambivalent or disagreed. Table 3 presents the results about the exercise beliefs and reported volume of exercise. The results show that the most highly correlated factors (beliefs) ($r > 0.2$) were the beliefs that exercise is important for long term management of the condition ($r = 0.341$, $p<0.001$), is important for control of pain ($r = 0.317$, $p < 0.001$), is important for well-being ($r = 0.296$, $p < 0.001$) and improves fitness ($r = 0.210$, $p < 0.001$). While Table 4 shows information on how reported symptoms correlate with reported exercise volume.

Table 5 provides the summary of the regression analysis results. Participants who received exercise advice from a physiotherapist were 1.8 times (80%) more likely to report a higher volume of weekly exercise (OR = 1.754 CI = 1.30-2.36, $p < 0.001$). Furthermore, participants who agreed with the belief that exercise is important for long term management of the condition were 2.8 times (280%) more likely to report that they undertook a high volume of weekly exercise compared to the participants who disagreed with this belief (OR = 2.76, 95% CI = 1.37-5.52, $p = 0.004$). Moreover, participants who agreed with the belief that exercise helps to control pain were 2.1 times (210%) more likely to undertake a high volume of exercise compared to the participants who strongly disagreed (OR = 2.11, 95% CI = 1.50-2.94, $p < 0.001$). Participants who agreed with the belief that exercise was important

for wellbeing were almost 1.7 (170%) times more likely to undertake a high volume of activities compared to the participants who disagreed (OR = 1.71, 95% CI = 1.05-2.79, p = 0.030). Those who were unemployed were 1.5 times (150%) less likely to undertake higher volumes of exercise than those who were in fulltime employment (OR = 0.538, 95% CI = 0.296-.976, p=0.041).

Exercise preferences and barriers to exercise

Participants were asked to list up to five types of exercise which they found most helpful. If exercise was unhelpful, they were asked to state 'exercise is unhelpful'. Swimming (n=267), walking (n=252) and Pilates (n=205) were the most frequently reported types of exercise. See Figure 2 for more detail.

Barriers to exercise were also explored. Eighty seven percent (n=824) of participants reported pain to be a barrier to exercise while fatigue 79% (n= 749) and fear of injury 50% (n = 475) were also commonly reported as barriers.

Experience of physiotherapy

A further aim of the research was to explore experiences of physiotherapy. Participants were given the opportunity to provide an open text answer to this question. The majority of participants, 81% (n=761) reported that they had received exercise advice from a physiotherapist 78% (n=746) had been given a prescription of exercise by a physiotherapist. Three themes emerged regarding experiences of physiotherapy.

Theme 1. Physiotherapist as a partner

Patients valued therapists who listened and worked in partnership to help them manage their condition. One female respondent who was sick listed at the time of completing the questionnaire and who was also diagnosed with co-existing fibromyalgia and who was now exercising for 2.5 hours per week states the following about her experience of recent physiotherapy.

"Only recently have I managed to find a physiotherapist who listens and is willing to work with me and help me to plan and manage my condition. It has made all the difference." P 32

Another female respondent who reported lower limb symptoms and migraines and who was also exercising for 2.5 hours or more per week stated the following about working with a physiotherapist with specialist training.

"My physiotherapist has specialised in hypermobility syndrome and has listened to what I have said concerning my body and what problems I have and helped construct me an exercise programme designed specifically to try and combat my worst problems - he has also listened when I've told him I am having problems with some exercises and told me when to stop them or has altered them for me so I can do them. So yes a great relationship of trust and understanding of both me as a person and my physical condition is what has made it work." P760

Theme 2. Communication, hand on guidance and feedback

Detailed explanations and feedback helps patients to understand how to do the exercises. For example one female respondent in the 18-20 year old age range, who was in full time employment and exercising 1-2.5 hours per week stated the following about the importance of explaining and 'hands on' guidance.

"It's really helpful when the physio explains, puts their hands on and shows you how to do the exercises when they give them to you, because you can't always tell if you're doing it right just from a line of text/static picture." P 767

The importance of guidance and feedback on exercises was also highlighted by a female respondent in the 31-40 years age group with lower back, knee and ankle symptoms.

"I found heavily guided exercise the most beneficial; I think that I am less likely to have awareness of how well I am completing the set tasks than "normal" people. My

last physio saw me for far longer than usual and also booked me follow up appointments monthly after each course finished so that she could keep checking my effectiveness of repetition afterwards, this enabled me to have plenty of feedback to keep my energy from being wasted by mis-performing exercises"' P759

Theme 3. Knowledge, experience and safety

The perceived effectiveness and safety of exercise was associated with therapist knowledge and experience. The following statement by one female respondent in the 41 – 50 year age range who suffered with widespread pain and symptoms for more than 20 years and who reported exercising for 2.5 hours per week or more.

"The specialist physio I am receiving is brilliant. Physio is useless though if you have a therapist who is inexperienced and knows nothing about hypermobility syndrome because they can prescribe exercises that can make you feel WORSE." P 620

Meanwhile another respondent, with co-existing Postural Tachycardia Syndrome, fibromyalgia and chronic fatigue, who was also exercising for 2.5 hours or more each week stated a similar sentiment.

"My first physio was diabolical, did no help whatsoever. Several years later I received more and was lucky enough to get someone who knew about hypermobility and started me off on Pilates and basic core physio work and it was excellent and made a big improvement to my quality of life. I'm now receiving physio on my shoulder by an EDS specialised physio and I've improved loads. It's just pot luck as to whether you get a physio with any knowledge of hypermobility as 'normal' physios are more harmful than beneficial."P38

Moreover a male respondent in the 31-40 year age group who suffered back pain, hip and knee pain, reported.

"The physiotherapist didn't seem to understand the condition. It was a waste of my time"P465

Discussion

The primary aim of this study was to explore the attitudes, beliefs and behaviours relating to exercise in the management of JHS/EDS-HT among adults living with the condition. Data from 946 questionnaires were analysed. It is not possible to report the response rate as the total sample frame of people with JHS/EDS-HT in both the patient organisations was unknown. Moreover many people are members of both the Hypermobility Syndromes Association (HMSA) and Ehlers Danlos Support UK (EDS UK) and therefore impossible to accurately establish the sample frame size. Respondent characteristics were similar to those reported by Rombaut and colleagues in 2011 in their study of 79 patients attending clinic in terms of age, gender, nature and duration of symptoms (13). Most participants were women and this is in line with the epidemiology (1-5). It should be noted that only 3 men offered comments in the open question relating to experiences of physiotherapy.

While the majority of participants recognised the general benefits of exercise, far fewer believed that it helped to control their pain. The influence of having received advice from a physiotherapist, beliefs that exercise is important for long term management, wellbeing and control of pain significantly impact on the reported exercise behaviour (i.e. volume of weekly exercise). Therapists should be aware that if patients are doubtful of the benefits of exercise in terms of symptom relief, it is unlikely that they will comply (31). Gecht et al., 1996 argued that patients' beliefs in the benefits of exercise are associated with exercise participation (36). In the systematic review conducted by Cooper et al. (2002), patients' views regarding exercise and current and past experience of exercise were important factors influencing attendance and adherence to cardiac rehabilitation programmes (37). Pain, fatigue and fear of injury were commonly reported barriers to exercise in this study. Similar

barriers have been reported amongst patients with other chronic conditions such as rheumatoid arthritis(38) and chronic back pain (39). A recent qualitative study relating to decision making amongst patients with JHS/EDS-HT attending pain management reported keeping pain at bay and at a manageable level was a feature of decision making process (40). A cost-benefit approach to decision making about activity which involved weighing the importance of an activity against its potential aversive consequences was used by the majority of individuals interviewed. Therapists therefore need to consider and discuss concerns and help them to weigh up the risks and benefits with patients when providing exercise advice.

While structured exercise such as Pilates and physiotherapy exercise were reported amongst the most helpful forms of exercise, therapists could consider offering a choice of exercise including other exercise such as swimming and walking as part of the exercise prescription. Patient choice has been shown to be an important factor for adherence with exercise amongst patients with chronic back pain (39). Moreover, aerobic exercise has been shown to be effective in managing symptoms of fibromyalgia (41), Postural Tachycardia Syndrome (42–45) and depression and anxiety(46). In particular swimming, walking and graded activity have been shown to be equally effective for improving pain and functional capacity in people with fibromyalgia (47). Given the known overlap between JHS/EDS-HT with fibromyalgia(48), depression and anxiety(49) and Postural Tachycardia Syndrome (17) these forms of exercise may be of significant benefit to this patient group.

Adherence and concordance with exercise and improved outcomes may be facilitated through the development of a positive therapeutic alliance(50). Health providers and in particular physiotherapists play a crucial role helping patients with chronic disorders such as JHS/EDS-HT to understand the nature of their disease, potential treatment benefits, addressing concerns regarding potential adverse effects and events and encouraging patients

to develop self-management and coping skills. Participants in this research study regarded communication, working in partnership with patients and being knowledgeable about the condition important and beneficial aspects of physiotherapy practice.

Limitations

This work was not without limitations. The recruitment from the Hypermobility Syndromes Association (HMSA) and Ehlers Danlos Support UK meant that the participants consisted of proactive people with an interest in their condition. People who agree to take part in research may also have different characteristics to those who do not. For example, the participants were largely highly educated individuals and may not represent the wider demographic. Furthermore the self-report method of questioning, this reveals a lot of information but this may not be correctly described which can make categorisation difficult, even with error. While the face validity of the questionnaire was developed with expert clinicians and patient representatives, full psychometric testing was not undertaken and this may impact on the validity and reliability of the results. To avoid ambiguity for participants, the term exercise was used in the research, even although technically, some activities which would normally be described as physical activity 30 when sets, repetitions, frequency are not included such as walking, swimming and cycling were reported as exercise. On the other hand, this study provides a substantial portrait of the patient population, albeit mainly highly educated women and the findings contribute data to understanding pertinent issues for people with JHS/EDS-HT engaging in exercise and physical activity. In the future research exploring the beliefs and behaviours of men and people with Hypermobility Spectrum Disorders from a broader education background should be considered.

Conclusions

Joint Hypermobility Syndrome and Ehlers Danlos Syndrome – Hypermobility Type are complex hereditary disorders of connective tissue and complex comorbidities may coexist.

The majority of individuals surveyed, believed exercise to be important in management. Individuals who received advice from a physiotherapist and the beliefs that exercise is important for long term management, wellbeing and control of pain significantly impacted on the reported exercise behaviour. Pain, fatigue and fear of injury were commonly reported barriers to exercise and physiotherapists should be mindful of these when advising and prescribing exercise. Verbal and non-verbal communication, working in partnership with patients and being knowledgeable about the condition are important and beneficial aspects of the therapeutic alliance and physiotherapy practice.

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Declaration of Interest

The authors report no declaration of interest

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Figure 1. Anatomic region affected

Figure 2. Types of exercise reported to be most helpful

Table 1: Demographic statistics in relation to the reported exercise behaviour (exercise volume)

		Reported exercise behaviour (volume of exercise per week)												Pearson Chi-square ($\chi 2$)	df	*p*-value
		I don't exercise		< 30 minutes		30-60 minutes		1-2.5 hours		> 2.5 hours		Total				
		Count	% of Total	Count	% of Total	Count	% of Total	Count	% of Total	Count	% of Total	Count	% of Total			
Gender	Female	96	10.1%	144	15.2%	188	19.9%	243	25.7%	235	24.8%	906	95.8%	1.94	4	0.748
	Male	5	0.5%	4	0.4%	9	1.0%	9	1.0%	13	1.4%	40	4.2%			
	Total	101	10.7%	148	15.6%	197	20.8%	252	26.6%	248	26.2%	946	100.0%			
Employment	Full time employed	23	2.4%	29	3.1%	54	5.7%	75	8.0%	72	7.6%	253	26.9%	14.45	8	<0.001
	Homemaker	7	0.7%	14	1.5%	15	1.6%	24	2.5%	18	1.9%	78	8.3%	.		
	Student	7	0.7%	22	2.3%	24	2.5%	40	4.2%	19	2.0%	112	11.9%			
	Sick listed	34	3.6%	49	5.2%	42	4.5%	54	5.7%	52	5.5%	231	24.5%			
	Unemployed	11	1.2%	8	0.8%	6	0.6%	6	0.6%	10	1.1%	41	4.4%			
	Retired	9	1.0%	6	0.6%	9	1.0%	6	0.6%	20	2.1%	50	5.3%			
	Part time employed	10	1.1%	20	2.1%	45	4.8%	46	4.9%	56	5.9%	177	18.8%			
	Total	101	10.7%	148	15.7%	195	20.7%	251	26.6%	247	26.2%	942	100.0%			
Marital status	Single	32	3.4%	54	5.7%	67	7.1%	98	10.4%	88	9.3%	339	35.9%	7.17	12	0.846
	Married / cohabiting	61	6.5%	86	9.1%	117	12.4%	137	14.5%	143	15.2%	544	57.7%			
	Separated / divorced	6	0.6%	6	0.6%	12	1.3%	14	1.5%	13	1.4%	51	5.4%			
	Widowed	0	0.0%	2	0.2%	0	0.0%	3	0.3%	4	0.4%	9	1.0%			
	Total	99	10.5%	148	15.7%	196	20.8%	252	26.7%	248	26.3%	943	100.0%			
Ethnicity	White	98	10.4%	142	15.0%	190	20.1%	242	25.6%	240	25.4%	912	96.4%	7.72	12	0.806
	Asian	1	0.1%	3	0.3%	2	0.2%	5	0.5%	1	0.1%	12	1.3%			
	Mixed	2	0.2%	2	0.2%	4	0.4%	2	0.2%	6	0.6%	16	1.7%			
	Other	0	0.0%	1	0.1%	1	0.1%	3	0.3%	1	0.1%	6	0.6%			
	Total	101	10.7%	148	15.6%	197	20.8%	252	26.6%	248	26.2%	946	100.0%			
Suffering time	> 20 years	56	5.9%	78	8.3%	87	9.2%	114	12.1%	138	14.6%	473	50.1%	20.48	16	0.199
	16 - 20 years	6	0.6%	14	1.5%	26	2.8%	29	3.1%	19	2.0%	94	10.0%			
	11 - 15 years	14	1.5%	29	3.1%	31	3.3%	42	4.4%	36	3.8%	152	16.1%			
	6 - 10 years	10	1.1%	14	1.5%	16	1.7%	20	2.1%	20	2.1%	80	8.5%			
	0 - 5 years	14	1.5%	13	1.4%	36	3.8%	47	5.0%	35	3.7%	145	15.4%			
	Total	100	10.6%	148	15.7%	196	20.8%	252	26.7%	248	26.3%	944	100.0%			
Exercise advice from PT	Yes	66	7.0%	109	11.5%	162	17.1%	216	22.8%	210	22.2%	763	80.7%	26.847	4	<0.001
	No	35	3.7%	39	4.1%	35	3.7%	36	3.8%	38	4.0%	183	19.3%			
	Total	101	10.7%	148	15.6%	197	20.8%	252	26.6%	248	26.2%	946	100.0%			

		Reported exercise behaviour (volume of exercise per week)												Spearman's rank Correlation Coefficient	*p*-value
		I don't exercise		< 30 minutes		30 - 60minutes		1 - 2.5 hours		> 2.5 hours		Total			
		Count	% of total	Count	% of total	Count	% of total	Count	% of total	Count	% of total	Count	% of total		
Age	18 - 20	13	1.4%	34	3.6%	37	3.9%	62	6.6%	35	3.7%	181	19.1%	0.05	.159
	21 - 30	19	2.0%	33	3.5%	68	7.2%	65	6.9%	57	6.0%	242	25.6%		
	31 - 40	33	3.5%	50	5.3%	51	5.4%	61	6.4%	64	6.8%	259	27.4%		
	41 - 50	21	2.2%	20	2.1%	22	2.3%	42	4.4%	55	5.8%	160	16.9%		
	51 - 65	11	1.2%	9	1.0%	14	1.5%	19	2.0%	33	3.5%	86	9.1%		
	65+	4	.4%	2	.2%	5	.5%	3	.3%	4	.4%	18	1.9%		
	Total	101	10.7%	148	15.6%	197	20.8%	252	26.6%	248	26.2%	946	100.0%		
Education	University / Further education	77	8.2%	116	12.4%	159	16.9%	194	20.7%	217	23.1%	763	81.3%	-0.08	.014
	Secondary education	23	2.4%	30	3.2%	34	3.6%	52	5.5%	31	3.3%	170	18.1%		
	Primary education	1	.1%	2	.2%	0	0.0%	3	.3%	0	0.0%	6	.6%		
	Total	101	10.8%	148	15.8%	193	20.6%	249	26.5%	248	26.4%	939	100.0%		

Table 2: Reported medical conditions

Medical condition	Number (%)
Mental health	**411 (43.4)**
Depression	259 (27.4)
Anxiety	85 (9)
Obsessive compulsive disorder	13 (1.3)
Other conditions ..	54 (5)
Musculoskeletal	**405 (42.8)**
Fibromyalgia	221(23.4)
Osteoarthritis	57 (6)
Scoliosis	26 (2.7)
Degenative spinal conditions	22 (2.3)
Other conditions..	79 (8)
Cardiorespiratory including cardiac dysautonomia	**385 (41)**
Postural Tachycardia Syndrome (Postural Tachycardia Syndrome)	200 (21)
Hypertension	74 (7.8)
Raynauds	46 (5)
Other conditions …	65 (7.2)
Gastrointestinal	**251 (27)**
Irritable Bowel Syndrome (IBS)	96 (10.1)
Gastro-Oesophageal Reflux Disorder (GORD)	42 (4.4)
Gastroparesis and dysmotility	29 (3.1)
Hiatus hernia	20 (2.1)
Other conditions….	64 (7.4)
Autoimmune	**223 (24)**
Asthma	128 (13.5)
Allergies	45 (5.1)
Other conditions …	50 (5.4)
Metabolic and nutritional	**120 (13)**
Hypothyroidism	35 (3.7)
Diabetes	19 (2.0)
Vit B12 deficiency	15 (1.6)
Other conditions ..	51 (5.7)
Fatigue and sleep related disorders	**99 (10)**
Chronic Fatigue Syndrome (CFS)	66 (7.0)
Insomnia	14 (1.5)
Other conditions …	19 (1.5)
Urogenital/ women's health	**97 (10)**
Poly Cystic Ovary Syndrome (PCOS)	29 (3.1)
Endometriosis	23 (2.4)
Other conditions …	45 (4.5)
Neurological/neurodevelopment	**35 (4)**
Arnold – Chiari Malformation	11 (1.2)
Asperger's Syndrome	8 (0.8)
Other conditions …	16 (1.5)

Table 3: Beliefs about exercise and reported exercise behaviour (volume)

Beliefs		**Reported exercise behaviour** (volume of reported exercises per week)												Spearman's rank Correlation Coefficient	*p*-value
		I don't exercise		< 30 minutes		30 - 60minutes		1 - 2.5 hours		➢ 2.5 hours		Total			
		Count	% of total	Count	% of total	Count	% of total	Count	% of total	Count	% of total	Count	% of total		
Exercise is important for long term management	Agree	51	5.4%	99	10.5%	144	15.2%	211	22.3%	219	23.1%	724	76.5%	-0.267	< 0.001
	Disagree	41	4.3%	42	4.4%	46	4.9%	36	3.8%	27	2.9%	192	20.3%		
	Undecided	9	1.0%	7	0.7%	7	0.7%	5	0.5%	2	0.2%	30	3.2%		
	Total	101	10.7%	148	15.6%	197	20.8%	252	26.6%	248	26.2%	946	100.0%		
Exercise is important for control of pain	Agree	18	19.0%	30	3.2%	71	7.5%	118	12.5%	156	16.5%	393	41.5%	-0.307	< 0.001
	Disagree	48	5.1%	67	7.1%	80	8.5%	83	8.8%	55	5.8%	333	35.2%		
	Undecided	35	3.7%	51	5.4%	46	4.9%	51	5.4%	37	3.9%	220	23.3%		
	Total	101	10.7%	148	15.6%	197	20.8%	252	26.6%	248	26.2%	946	100.0%		
Exercise is important for wellbeing	Agree	59	6.2%	102	10.8%	155	16.4%	208	22.0%	217	22.9%	741	78.3%	-0.207	< 0.001
	Disagree	25	2.6%	34	3.6%	28	3.0%	31	3.3%	20	2.1%	138	14.6%		
	Undecided	17	1.8%	12	1.3%	14	1.5%	13	1.4%	11	1.2%	67	7.1%		
	Total	101	10.7%	148	15.6%	197	20.8%	252	26.6%	248	26.2%	946	100.0%		
Exercise improves fitness	Agree	83	8.8%	126	13.4%	180	19.1%	231	24.5%	230	24.4%	850	90.3%	-0.108	0.001
	Disagree	8	.9%	16	1.7%	10	1.1%	17	1.8%	10	1.1%	61	6.5%		
	Undecided	10	1.1%	4	0.4%	6	0.6%	4	.4%	6	0.6%	30	3.2%		
	Total	101	10.7%	146	15.5%	196	20.8%	252	26.8%	246	26.1%	941	100.0%		
Exercise helps control weight	Agree	79	8.4%	118	12.5%	168	17.9%	214	22.7%	211	22.4%	790	84.0%	-0.047	0.150
	Disagree	13	1.4%	21	2.2%	18	1.9%	29	3.1%	20	2.1%	101	10.7%		
	Undecided	8	00.9%	7	0.7%	10	1.1%	9	1.0%	16	1.7%	50	5.3%		
	Total	100	10.6%	146	15.5%	196	20.8%	252	26.8%	247	26.2%	941	100.0%		
Exercise mental alertness	Agree	51	5.4%	62	6.6%	82	8.7%	107	11.4%	79	8.4%	381	40.4%	0.174	< 0.001
	Disagree	31	3.3%	52	5.5%	55	5.8%	63	6.7%	48	5.1%	249	26.4%		
	Undecided	17	1.8%	34	3.6%	58	6.2%	82	8.7%	121	12.8%	312	33.1%		
	Total	99	10.5%	148	15.7%	195	20.7%	252	26.8%	248	26.3%	942	100.0%		

Table 4: Association between exercise behaviour and commonly reported symptoms

		Exercise Behaviour (volume of reported exercise per week)												Spearman's rank Correlation Coefficient	*p*-value
		I don't exercise		< 30 minutes		30 - 60minutes		1 - 2.5 hours		> 2.5 hours		Total			
Symptoms		Count	% of total	Count	% of total	Count	% of total	Count	% of total	Count	% of total	Count	% of total		
Joint pain	Never-Rarely	1	0.1%	0	0.0%	3	0.3%	1	0.1%	1	0.1%	6	0.6%	-0.021	0.511
	Sometimes	4	0.4%	7	0.7%	7	0.7%	10	1.1%	15	1.6%	43	4.5%		
	Frequently	96	10.1%	141	14.9%	187	19.8%	241	25.5%	232	24.5%	897	94.8%		
	Total	101	10.7%	148	15.6%	197	20.8%	252	26.6%	248	26.2%	946	100.0%		
Joint stiffness	Never-Rarely	6	0.6%	6	.6%	27	2.9%	23	2.4%	21	2.2%	83	8.8%	-0.059	0.068
	Sometimes	12	1.3%	18	1.9%	31	3.3%	44	4.7%	42	4.4%	147	15.5%		
	Frequently	83	8.8%	124	13.1%	139	14.7%	185	19.6%	185	19.6%	716	75.7%		
	Total	101	10.7%	148	15.6%	197	20.8%	252	26.6%	248	26.2%	946	100.0%		
Tender point	Never-Rarely	3	0.3%	0	0.0%	7	0.7%	7	0.7%	8	25	0.8%	2.6%	-0.056	0.087
	Sometimes	8	0.8%	16	1.7%	28	3.0%	31	3.3%	35	3.7%	118	12.5%		
	Frequently	90	9.5%	132	14.0%	162	17.1%	214	22.6%	205	21.7%	803	84.9%		
	Total	101	10.7%	148	15.6%	197	20.8%	252	26.6%	248	26.2%	946	100.0%		
Muscle stiffness	Never-Rarely	8	0.8%	4	0.4%	20	2.1%	23	2.4%	31	3.3%	86	9.1%	-0.062	0.058
	Sometimes	21	2.2%	29	3.1%	57	6.0%	69	7.3%	51	5.4%	227	24.0%		
	Frequently	72	7.6%	115	12.2%	120	12.7%	160	16.9%	166	17.5%	633	66.9%		
	Total	101	10.7%	148	15.6%	197	20.8%	252	26.6%	248	26.2%	946	100.0%		
Muscle spasm	Never-Rarely	16	1.7%	15	1.6%	37	3.9%	42	4.4%	41	4.3%	151	16.0%	-0.034	0.294
	Sometimes	33	3.5%	44	4.7%	78	8.2%	99	10.5%	81	8.6%	335	35.4%		
	Frequently	52	5.5%	89	9.4%	82	8.7%	111	11.7%	126	13.3%	460	48.6%		
	Total	101	10.7%	148	15.6%	197	20.8%	252	26.6%	248	26.2%	946	100.0%		
Poor balance	Never-Rarely	6	0.6%	4	0.4%	14	1.5%	30	3.2%	28	3.0%	82	8.7%	-0.154	< 0.001
	Sometimes	18	1.9%	37	3.9%	54	5.7%	86	9.1%	79	8.4%	274	29.0%		
	Frequently	77	8.1%	107	11.3%	129	13.6%	136	14.4%	141	14.9%	590	62.4%		
	Total	101	10.7%	148	15.6%	197	20.8%	252	26.6%	248	26.2%	946	100.0%		
Joint subluxations and dislocations	Never-Rarely	19	2.0%	37	3.9%	55	5.8%	81	8.6%	61	6.4%	253	26.7%	-0.049	0.124
	Sometimes	26	2.7%	43	4.5%	50	5.3%	63	6.7%	78	8.2%	260	27.5%		
	Frequently	56	5.9%	68	7.2%	92	9.7%	108	11.4%	109	11.5%	433	45.8%		
	Total	101	10.7%	148	15.6%	197	20.8%	252	26.6%	248	26.2%	946	100.0%		
Bruising	Never-Rarely	11	1.2%	18	1.9%	23	2.4%	45	4.8%	40	4.2%	137	14.5%	-0.061	0.061
	Sometimes	32	3.4%	56	5.9%	62	6.6%	81	8.6%	88	9.3%	319	33.7%		
	Frequently	58	6.1%	74	7.8%	112	11.8%	126	13.3%	120	12.7%	490	51.8%		
	Total	101	10.7%	148	15.6%	197	20.8%	252	26.6%	248	26.2%	946	100.0%		
Dizziness / fainting	Never-Rarely	12	1.3%	21	2.2%	26	2.8%	40	4.2%	39	4.1%	138	14.6%	-0.105	0.001
	Sometimes	29	3.1%	50	5.3%	83	8.8%	104	11.0%	109	11.6%	375	39.8%		
	Frequently	60	6.4%	77	8.2%	88	9.3%	106	11.3%	98	10.4%	429	45.5%		
	Total	101	10.7%	148	15.7%	197	20.9%	250	26.5%	246	26.1%	942	100.0%		
Bowel symptoms	Never-Rarely	21	2.2%	28	3.0%	46	4.9%	76	8.0%	57	6.0%	228	24.1%	-0.059	.071
	Sometimes	18	1.9%	31	3.3%	57	6.0%	58	6.1%	61	6.4%	225	23.8%		
	Frequently	62	6.6%	89	9.4%	94	9.9%	118	12.5%	130	13.7%	493	52.1%		
	Total	101	10.7%	148	15.6%	197	20.8%	252	26.6%	248	26.2%	946	100.0%		
Fatigue	Never-Rarely	1	0.1%	1	0.1%	1	0.1%	0	0.0%	6	0.6%	9	1.0%	-0.12	< .001
	Sometimes	6	0.6%	9	1.0%	28	3.0%	34	3.6%	41	4.3%	118	12.5%		
	Frequently	94	9.9%	138	14.6%	168	17.8%	218	23.0%	201	21.2%	819	86.6%		
	Total	101	10.7%	148	15.6%	197	20.8%	252	26.6%	248	26.2%	946	100.0%		

Table 5: Model Fitting Information

Model	-2 Log Likelihood	$\chi 2$	df	p-value
Intercept Only	2105.688			
Final	1905.702	199.986	17	< 0.001

Pseudo R-Square

Cox and Snell	0.192
Nagelkerke	0.201
McFadden	0.068

Goodness-of-Fit

	$\chi 2$	df	p-value
Pearson	1775.726	1715	0.150
Deviance	1506.235	1715	1

Test of Parallel Lines

Model	-2 Log Likelihood	$\chi 2$	df	p-value
Null Hypothesis	1905.7			
General	1837.5	68.192	51	0.054

		Estimate	Std. Error	Wald	df	p-value	95% Confidence Interval		Odd ratio	95% Confidence Interval	
							Lower Bound	Upper Bound		Lower Bound	Upper Bound
Volume of exercise	No exercise - < 30 minutes	-0.688	0.412	2.781	1	0.095	-1.496	0.121	0.503	0.224	1.128
	< 30 minutes - 30 -60 mins	0.563	0.411	1.876	1	0.171	-0.242	1.368	1.756	0.785	3.928
	30-60 mins - 1-2.5 hours	1.620	0.414	15.292	1	<0 .001	0.808	2.431	5.051	2.243	11.373
	1-2.5 hours - > 2.5 hours	2.946	0.420	49.165	1	< .001	2.123	3.770	19.038	8.355	43.383
Advice from physiotherapist	Yes	.562	.152	13.710	1	< .001	.264	.859	1.754	1.303	2.362
	No	0							1		
Employment status	Part time employed	0.173	0.179	0.930	1	0.335	-0.179	0.524	1.189	0.836	1.690
	Homemaker	-0.029	0.236	0.015	1	0.902	-0.491	0.433	0.971	0.612	1.542
	Student	0.014	0.208	0.005	1	0.946	-0.394	0.423	1.014	0.674	1.526
	Sick listed	-0.250	0.170	2.167	1	0.141	-0.582	0.083	0.779	0.559	1.086
	Unemployed	-0.620	0.304	4.160	1	0.041	-1.216	-0.024	0.538	0.296	0.976
	Retired	0.175	0.287	0.372	1	0.542	-0.388	0.738	1.192	0.678	2.093
	Full time employed	0							1		
Poor balance	Never-Rarely	0.412	0.222	3.435	1	0.064	-0.024	0.848	1.510	0.977	2.335
	Sometimes	0.372	0.137	7.350	1	0.007	0.103	0.642	1.451	1.109	1.899
	Frequently	0							1		
Exercise long term management	Agree	1.013	0.355	8.137	1	00.004	0.317	1.710	2.755	1.373	5.527
	Undecided	0.388	0.362	1.145	1	0.285	-0.322	1.098	1.473	.724	2.997
	Disagree	0							1		
Exercise control pain	Agree	0.745	0.172	18.799	1	0.000	0.408	1.082	2.106	1.504	2.950
	Undecided	-0.169	0.163	1.068	1	0.301	-0.489	0.151	0.845	0.613	1.163
	Disagree	0							1		
Exercise wellbeing	Agree	0.538	0.248	4.694	1	0.030	0.051	1.025	1.713	1.053	2.786
	Undecided	0.230	0.281	0.669	1	0.414	-0.321	0.780	1.258	0.726	2.181
	Disagree	0							1		
Exercise mental	Agree	-0.678	0.144	22.221	1	< 0.001	-0.960	-0.396	0.507	0.383	0.673
	Undecided	-0.422	0.166	6.457	1	0.011	-0.748	-0.097	0.656	0.473	0.908
	Disagree	0							1		

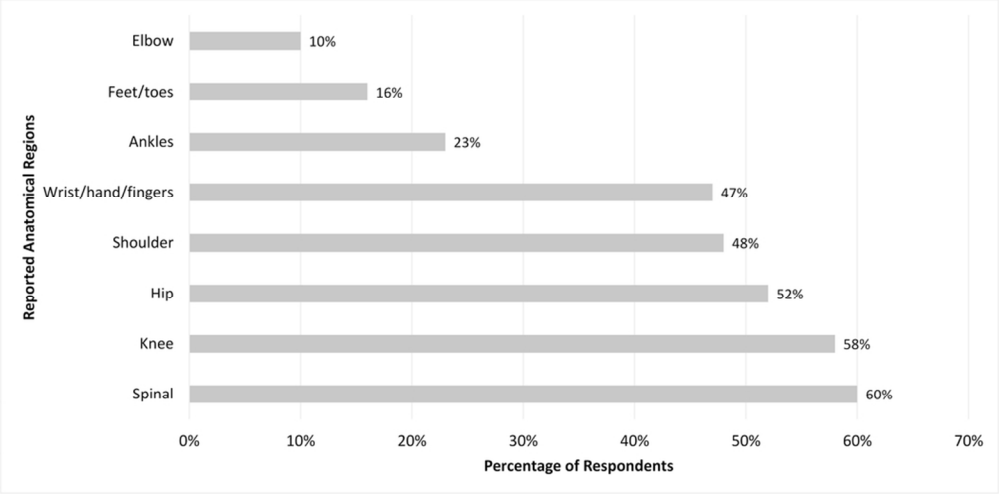

Figure 1. Most problematic region of the body

89x44mm (300 x 300 DPI)

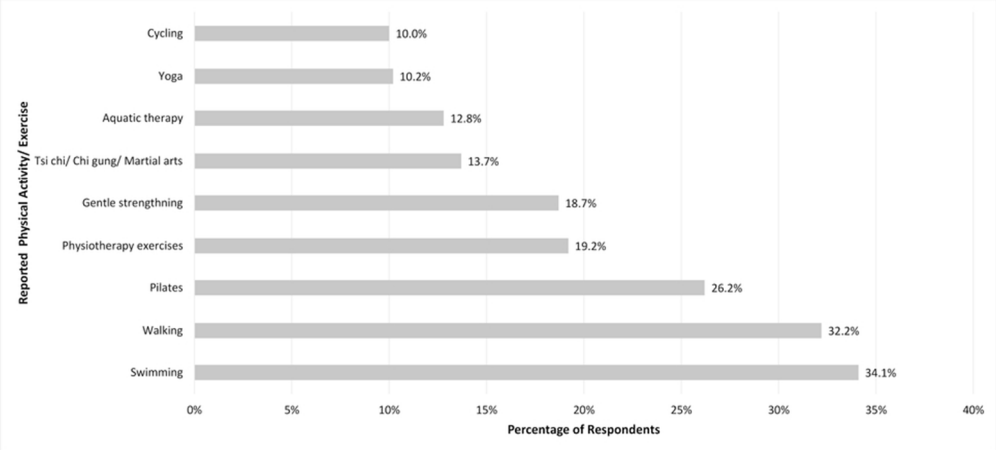

Figure 2. Types of exercise reported to be most helpful

81x37mm (300 x 300 DPI)

Implications for Rehabilitation

- Exercise is a cornerstone of treatment for Ehlers Danlos Syndrome/ Ehlers Danlos Syndrome – Hypermobility Type
- Pain, fatigue and fear of injury are frequently reported barriers to exercise
- Advice from a physiotherapists may significantly influence exercise behaviour
- Physiotherapists with condition specific knowledge and good verbal and non verbal communication facilitate a positive therapeutic experience

Exercise and Joint Hypermobility Questionnaire

This questionnaire aims to explore your experience and beliefs and about exercise. Please answer all the questions. For most questions, please make a mark (X) the appropriate box answer. For some questions you are asked to write a short answer.

SECTION 1 Demographic and clinical information

1	**What is your age?**		
	18-20		
	21-30		
	31 -40		
	41-50		
	51 - 65		
	65 or older		
2.	**What gender are you?**		
	Male		
	female		
3.	**What is your ethnicity?**		
	White		
	Asian - Pakistan		
	Asian - Indian		
	Asian - Chinese		
	Asian - other		
	Mixed		
	Black African		
	Black Caribbean		
	Black other		
	0ther		
4.	**What is your marital status?**		
	Married/ cohabiting/ civil partnership		
	Single		
	Divorced		
	Widowed		
5.	**Highest education level achieved?**		
	Primary school		
	Secondary school		
	University or further education		
6.	**Which of the following best describes your current employment status?**		
	Full time (30 hours or more)		
	Part time		
	Home maker		
	Unemployed		
	Sick listed		
	Retired		
	Student		
7.	**Which condition/s have you been diagnosed with?**		
	Joint Hypermobility Syndrome (JHS)		
	Ehlers Danlos Syndrome - Hypermobility Type (EDS-HT)		
	Both JHS and EDS-HT		

8.	**How long have you suffered from symptoms of JHS/EDS-HT?**					
	0-4 years					
	5-10 years					
	11-20 years					
	More than 20 years					
9.	**During the past 3 months, which parts of your body have given you the most problems? List and rank most problematic regions. For example 1. Shoulder, 2. Knee, 3. Ankle**					
	1.					
	2.					
	3.					
	4.					
	5.					
10.	*During the past 3 months, how often have you experienced the following?*					
		Never	Rarely	Sometimes	Frequently	Constant
	Joint pain					
	Joint stiffness					
	Muscle spasm					
	Poor balance					
	Fatigue					
	Bruising					
	Dizziness/ feeling faint					
	Joint subluxation/ dislocation					
	Irritable bowel symptoms					
	Poor balance					
11.	**Who first diagnosed your JHS or EDS-HT?**					
	General practitioner					
	Rheumatologist					
	Physiotherapist					
	Other					
12.	**Do you suffer from any other medical conditions (eg high or low blood pressure, depression, arthritis etc)**					
	Yes					
	No					
13.	**If you answered yes to question 12, please state which condition/s**					

SECTION 2 Exercise Beliefs					
14.	Have you received exercise advice from a physiotherapist for JHS/ EDS-HT?				
	Yes				
	No				
15.	**What types of exercise have you found most helpful? Please list up to 5 of the most helpful types of exercise. If exercise is not helpful please state, exercise is not helpful.**				
	1.				
	2.				
	3.				
	4.				
	5.				
16.	**Please state your strength of agreement with the following statements**				
	Strongly agree	Agree	Undecided	Disagree	Strongly disagree
Exercise is important for long term management of JHS/ EDS-HT					
Exercise is important for control of pain					
Exercise is important for wellbeing					
Exercise improves sleep					
Exercise improves fitness					
Exercise helps control weight					
Exercise improves mental alertness					

SECTION 3 Exercise Behaviours and Barriers			
17.	**On average during the past 3 months, how long have you spent exercising each week**		
	1. I don't exercise		
	2. Less than 30 minutes		
	3. Between 30 and 60 minutes		
	4. Between 1 and 2.15 hours		
	5. More than 2.5 hours		
18.	**Please indicate which of the following prevent you from exercising (Please cross all that apply)**		
	1. Not sure which exercise to do		
	2. Lack of time		
	3. Pain		
	4. Fear of injury		
	5. Fatigue		
	6. Embarrasment		
	7. Lack of motivation		
	8. Weather		
	9. No one to exercise		
	10. Other (please specify)		
19.	**What is the likelihood of you feeling able to exercise in the following situations? (where 1 is the most likely and 10 is the least likely)**		
	1.When feeling tired		
	2.When my joints and muscles ache		
	3. When under time pressure		
	4.When feeling anxious		
	5.When I have too many other things to do		
	6. After recovering from an injury		
	7.When on holiday		
	8. When returning from a holiday		
	9.When the weather is bad		
	10.When I do not have the support of my family or friends		
SECTION 4. Experiences of Physiotherapy			
20.	Have you been prescribed exercise/s by a physiotherapist?		
	Yes		
	No		
21.	If you answered yes to question 20, in order for us to improve our services, please describe your experience of physiotherapy.		