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Subjective health complaints and illness perception amongst adults with Joint hypermobility syndrome/Ehlers Danlos Syndrome - hypermobility type - a cross-sectional study.

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Abstract:

Objective: To investigate the prevalence and severity of subjective health complaints and describe illness perception in a population of Joint Hypermobility Syndrome or Ehlers Danlos Syndrome-Hypermobile Type.

Method: This study was a postal survey with a questionnaire battery on demographic data, subjective health complaints inventory, and illness perception. A total of 110 individuals diagnosed with Joint Hypermobility Syndrome or Ehlers Danlos Syndrome-Hypermobile Type from two specialized hospitals in Norway were offered participation. Further, 140

gender- and age-matched healthy controls from statistics Norway representing the general population were sent the questionnaire for reference.

Results: Overall response rate was 30.4% (n=76), with 44.5% (n=49) in Joint Hypermobility Syndrome/ Ehlers Danlos Syndrome-Hypermobile Type and 19.3% (n=27) in controls. Subjective health complaints were significantly higher in Joint Hypermobility Syndrome/ Ehlers Danlos Syndrome-Hypermobile Type - than in controls (32.06 vs 11.08; $p < 0.001$). Further the brief illness perception questionnaire indicated that the adults with Joint Hypermobility Syndrome/ Ehlers Danlos Syndrome-Hypermobile Type had low understanding of their illness and symptoms (understanding, mean: 3.93, SD 2.88), and reported to have moderate personal and treatment control over their illness.

Conclusion: Adults with Joint Hypermobility Syndrome/ Ehlers Danlos Syndrome-Hypermobile Type reported higher frequency and severity of subjective health complaints than matched controls from the general adult population in Norway. Furthermore, Joint Hypermobility Syndrome/ Ehlers Danlos Syndrome-Hypermobile Type reported low understanding of their illness and associated symptoms, and moderate belief that their illness can be kept under control through self-management or treatment. This may indicate one of the reasons why prognosis for these patients is poor.

Keywords

Joint Instability • Surveys and Questionnaires • Illness perceptions • Diagnostic Self Evaluation. •Subjective health complaints

INTRODUCTION

Generalized Joint Hypermobility (GJH) is a trait commonly seen in healthy individuals with no health complaints (1). Joint hypermobility refers to the ability to extend one or more joints beyond what is considered normal (2). When joint hypermobility is occurring in multiple joints it is classified as GJH (3). When GJH becomes symptomatic it is often referred to as Joint Hypermobility Syndrome (JHS) (2). The estimated prevalence of JHS varies from 2-57% (1). Symptomatic GJH (JHS) is associated with functional and psychosocial impairment, manifesting with a variety of musculoskeletal and visceral complaints, and most often diminished health-related quality of life (4-6). GJH is a dominant trait seen in several Heritable Connective Tissue Disorders, including Ehlers–Danlos Syndrome Hypermobile Type (EDS-HT) (2). Due to similarities in the clinical presentation and a significant phenotypic overlap of these syndromes, an international panel of experts has recommended EDS-HT as indistinguishable from JHS (3, 7). Hence, the two terms JHS/EDS-HT are for the purpose of this study considered inter-changeable.

In the last few years numerous articles on 'non'-musculoskeletal problems in the JHS/EDS-HT population, such as fatigue, orthostatic intolerance and gastrointestinal complaints have been published (5, 6, 8). Other studies have further reported that a variety of health complaints are both common and substantial in the JHS/EDS-HT population (9, 10). Musculoskeletal pain, fatigue, dizziness, headache and mood swings are common complaints also in the general population (11). Totally 96% of the general population in Norway report at least one health complaint within the latest 30 days (11, 12). These multiple common health complaints are often described as "subjective health complaints" (SHC) (11), and cover five major subgroups; musculoskeletal complaints, gastrointestinal complaints, pseudo-neurological complaints, symptoms of allergies and cold (11). SHCs are the leading cause of

long-term sickness and disability (12). Several studies have suggested that sensitization may be a psycho-biological mechanism that could play a role in the development of chronic and severe SHC (13-16).

The Self-Regulation Model is frequently used as a framework for describing the perceptions people have about their health condition (17). The Self-Regulation Model proposes that individuals create common-sense beliefs regarding their illness, and that these beliefs are important for understanding and coping with perceived health threats (18). These beliefs are frequently referred to as illness perceptions and may be characterized as adaptive or maladaptive (19).

Leventhal and colleagues (17) originally describe five dimensions of these illness representations: *identity*– the symptoms attributed to the illness and the label used to describe it, *consequences* beliefs about the severity of the illness and how it will impact on life, *timeline*- expectations about how long the illness will last, *control/cure* - whether the illness can be cured or controlled by self-management or treatment and *cause*- personal thoughts about the cause of the illness. Further, *understanding* - the patient's sense of coherence with regard to the illness in question, and *emotional representations* - the patients emotional response to the illness and its symptoms, where included later as additional dimensions in illness perceptions (17).

The relation between illness perceptions and health-related outcomes has been widely studied in several patient populations, and illness perceptions are generally accepted as generically important to patients´ health behaviors and -outcomes (18, 20). Furthermore, illness perceptions have been shown to influence treatment adherence and functional recovery (20-22). In general, adaptive illness perceptions guide a person towards health behaviors that have a positive contribution to health, while maladaptive illness perceptions may guide a person

toward negative, inexpedient health behaviors (21, 23). In addition to the associations with behavioral and quality-of-life outcomes, there is growing evidence that severe health complaints are linked to the behavior of the individual E.g. highly fearful persons who tend to have catastrophizing thoughts will avoid activities they perceive as harmful or pain provoking. In the long term, this avoidance behavior can result in physical inactivity, disability and depression, further fueling the vicious circle of disabling musculoskeletal pain (24). As far as we know, there have been no previous attempts to explore illness perceptions in JHS/EDS-HT patients.

Hence, this study has three aims: i) to investigate the prevalence and severity of subjective health complaints in Norwegian adults with JHS/EDS-HT, ii) to compare these results with a matched control group representing a general adult population, (iii) and to create a profile of illness perceptions in adults with JHS/EDS-HT.

METHODS AND MATERIALS

Procedures

This study is part of a study reporting shoulder specific problems in this group (25). The study was conducted as a postal survey with a questionnaire battery including demographic data, subjective health complaints inventory (SHC-I), and illness perceptions (B-IPQ). The participants with JHS/EDS-HT (cases) were recruited based on availability from Haukeland University Hospital, Department of physical medicine and rehabilitation, Bergen, Norway, and the Training and counseling center for rare disorders, Sunnaas rehabilitation hospital, Nesodden, Norway.

An age and gender matched control group was randomly selected from Statistics Norway (called controls). In total, the questionnaires were sent out by mail to 250 individuals (110 individuals with JHS/EDS-HT and 140 controls) with a three- week deadline. After the deadline a reminder was sent out by mail. All participants responded in the time period between February and April 2014.

Inclusion and Exclusion Criteria

Men and women between the ages of 16-67 were included in the study. Due to the relative low number of individuals with JHS/EDS-HT, the exclusion criteria for both cases and controls were few. Participants were only excluded if they had been pregnant or given birth within the latest year, if they had additional neurological or rheumatic diseases, or were in lack of a confirmative diagnosis of JHS/EDS-HT.

ETHICAL CONSIDERATIONS

This study was approved by the Regional Committee for Medical and Health Research Ethics, Norway (Ref. no. 2013/2011). Participants were informed of the voluntary nature of participating, and informed consent to participate according to the Declaration of Helsinki was assumed based on the return of the questionnaires.

Methods/Questionnaires

The SHC- inventory

SHC was measured with a standardized instrument, the "Subjective health complaints inventory" (SHC-I) (26). The inventory registers subjective reports of incidence, in addition to extent of health complaints during the past 30 days, as related to 29 common somatic and

mental health complaints. The self-rated levels of affect are graded on a four-point Likert scale: 0 = not bothered, 1 = slightly bothered, 2 = partly bothered and 3 = severely bothered. The 29 individual complaints are commonly reported on a total score (SHC-total), but can also be grouped into five subscales (26): *Musculoskeletal complaints* (headache, neck pain, upper back pain, low back pain, arm pain, shoulder pain, feet pain, migraine), *pseudo neurological complaints* (palpitations, hot flashes, sleep problems, fatigue, dizziness, anxiety, depression), *gastrointestinal complaints* (heartburn, gas discomfort, diarrhoea, obstipation, abdominal pain, ulcer and bellyache), *allergic complaints* (asthma, allergies, breathing difficulties, chest pain and eczema), *and colds* (colds, flu, coughs, bronchitis). The SHC inventory has demonstrated good reliability (26).

The Brief illness perception questionnaire (B-IPQ)

The B-IPQ consists of nine items, of which the first eight items are (1) consequences, (2) timeline, (3) personal control, (4) treatment control, (5) identity, (6) concern, (7) coherence and (8) emotional representation (27). Reponses to these eight items are given on an eleven point response scale ranging from 0 - 10. The B-IPQ also includes a ninth, open item asking participants to list the three most important causes of their illness or health complaints. The B-IPQ has demonstrated good test–re-test reliability and validity (18, 27, 28). A higher score reflects a more threatening view of the illness (28). However, there is no consensus on specific cut off points relating to the degree of which the illness is perceived as threatening or benign, since the cut off and internal consistency will depend on the illness population (28).

Demographics

Socio-demographic indices including gender, height, weight, education, activity level, employment status, and questions concerning subjective assessment of participants' health and work ability, were further collected from the questionnaire battery.

STATISTICAL ANALYSES

Data was tested for normality by Shapiro-Wilks. Demographics (gender, age and educational level) were presented by descriptive analyses (frequency, mean and SD).
In addition to the total score for each of the five subscales of SHC, a mean score was calculated (38). For each of the eight items of B-IPQ a mean group value was calculated. An overall score was computed by reversing the scores of items 3, 4, and 7 and adding them to items 1, 2, 5, 6, and 8 in the total score (27).

Group differences on SHC were tested by independent sample t-test, one-way ANOVA for parametric data, and with Chi2 and Mann-Whitney's U-test for non-parametric data.

SPSS version 23 for Windows was used for the statistical analyses, and level of significance was selected as $p \leq 0.05$.

RESULTS

Demographic data

The characteristics of the participants are shown in table I. In total 76 individuals returned the questionnaire, corresponding to total response rate 30.4%, with n= 49 (44.5%) for cases, and n= 27 (19.3%) for controls (Figure I).

The average age was 40.6 years (+ 12.6), and 93.4 % (n=71) of the participants were women. The two samples did not differ significantly with respect to gender, age, BMI, educational level and employment status (Table I). More of the JHS/EDS-HT participants were on 'sick leave/disability pension' and 'employed and partially on disability pension' compared with controls (Table I).

Subjective health complaints (SHC)

All (100%) participants in the JHS/EDS-HT group reported having experienced at least one musculoskeletal complaint during the latest 30 days (Figure II). The most frequent complaints among JHS/EDS-HT were headache, neck pain, tiredness, sleep problems, dizziness, arm pain, foot pain, shoulder pain, stomach ache, gas discomfort, upper and lower back pain, reported by 74% to 96% of the JHS/EDS-HT participants (Figure II). In the matched control group, 84% reported having at least one musculoskeletal complaint during the past 30 days. The most common single complaint in controls was neck pain (46%), gas discomfort (46%), headaches (50%) and tiredness (53%).

The self-reported level of SHC severity (SHC total score) was significantly higher in JHS/EDS-HT than in controls (32.06 vs 11.08; $p < 0.001$) (Table II). On the SHC subscales JHS/EDS-HT reported significantly more complaints than the control group on musculoskeletal ($p<0.001$; except for migraine), pseudo-neurological ($p < 0.001$; except from anxiety), gastrointestinal ($p< 0.001$; except from ulcer) and allergic ($p= 0.001$; except from chest pain). There were no differences in flu/cold complaints.

Illness perceptions

Overall, the participants with JHS/EDS-HT perceived their illness to be chronic (timeline, mean: 9.39) (Table III) and reported that their illness was comprehensive, indicating that they had low understanding of their illness and symptoms (understanding, mean: 3.93). There was a general perception of having moderate personal control of the illness (mean: 5.39) and moderate control of the illness through treatment (mean: 4.98). The mean total score for the B-IPQ was 47.17.

DISCUSSION

This study found a high prevalence of SHC amongst this sample of adults with JHS/EDS-HT. As many as 100 % of the sample had experienced at least one complaint during the past month, and the severity of the total, and SHC subscales, were up to three times higher in JHS/EDS-HT compared with controls. The JHS/EDS-HT sample perceived their illness to be chronic, they reported low understanding and comprehension of their illness and symptoms, and expressed pessimism about the role treatment could have in controlling their symptoms.

In addition to musculoskeletal pain, the majority of the individuals with JHS/EDS-HT reported a high level of pseudoneurological and gastrointestinal complaints. These results are in accordance with previous studies (6, 10, 29) and strengthens the argument that JHS/EDS-HT may be defined as a generalized disorder tagging involvement of a variety of organ systems, such as the gastrointestinal (30) and neuromuscular systems (31).

Grahame et al. (45) suggested that pain in JHS/EDS-HT was directly linked to primary joint damage and, therefore mainly the pain was indicative of nociceptive origin (32). However, a pilot study by Camerota et al. (33) suggested that pain in EDS-HT may also have neuropathic origin. Recent work by Rombaut et al. (34) described the presence of generalized hyperalgesia in adults with EDS-HT resulting in lower pressure thresholds in both symptomatic and asymptomatic areas, compared with healthy controls. The same was demonstrated in individuals with JHS (60). Continuous exposure to pain may lead to a sensitization process as a result of neuronal hyper excitability involving both peripheral and central structures of the nociceptive system. Central sensitization is understood as neuronal hyper excitability in response to peripheral stimuli that permanently modifies sensory processes (16, 35). Central sensitization of nociceptive stimuli has been suggested as a mechanism for understanding several syndromes with high levels of pain and comorbid health complaints (36, 37).

Neurophysiological changes compatible with central sensitization are now recognized in understanding chronic pain in rheumatoid conditions, such as rheumatoid arthritis, osteoarthritis and fibromyalgia (38). Several studies have found EDS to have close resemblance with fibromyalgia as both conditions are characterized by pain, fatigue and physical disability (4, 9). Females with EDS rated their functional health status as worse than females with fibromyalgia (48). Knowledge of these high levels of disabling health complaints in this population is recommended to have consequences for management and choice of treatment strategies chosen by health care professionals.

With regard to illness perceptions, the present study found low scores on the B-IPQ dimension *coherence* (understanding), assessing whether the illness "makes sense" for the patient. In other words, our study population of JHS/EDS-HT reports a lack of understanding of their disease and symptoms. This finding is in line with a recent study where the symptoms of JHS/EDS- HT were described as unpredictable, diverse, and fluctuating in nature (39). Healthcare professionals should take this into consideration in the rehabilitation process, since high sense of coherence in relation to one's health complaints, is important for long-term, positive emotional health outcomes in individuals with chronic illness (40). Further, patients need to understand the nature of their symptoms or illness to be able to adopt beneficial coping strategies (41). The high variability in the clinical presentations and the lack of visible signs and symptoms add to the complexity of JHS/EDS-HT and makes it difficult for both patients' and health care providers to deal with these conditions. The consequence may be that JHS/EDS-HT patients and their health problems are under-recognized and inadequately managed by health professionals (8) in the rehabilitation process. This adds burden to the patients and may explain the present findings that they don't understand their own disease. Lack of understanding may also cause patients to develop avoiding strategies, such as

kinesiophobia (42), requiring further exploration of health professionals understanding and treatment strategies for this group.

The participants in our study reported moderate sense of *personal control* over their illness, and appeared less optimistic about the role treatment could have in controlling their symptoms. The personal treatment control dimension of B-IPQ refers to the feeling of empowerment of effective coping behavior (20). Moreover, studies have shown that having a sense of personal control and a confidence in treatment, results in higher treatment adherence in patients with diabetes and hypertension (43, 44). Leading to secondary benefits such as better social functioning and well-being, lower distress and higher vitality (45).

There is growing evidence that illness perceptions are modifiable (46), as several randomized controlled trials have shown that the dimension of *personal control* changes most easily in response to interventions including cognitive behavioral therapy, educational programs and psycho-education programs (40). Providing structured patient education has shown to be an essential component in managing a wide range of chronic illnesses, as described recently in an uncontrolled study on EDS-HT (47). Further, in patients with rheumatoid arthritis, patient education was shown to have an immediate and beneficial effect on self-reported factors such as disability, psychological status, and depression (48). As this study cannot give clear recommendations regarding content and efficacy of patient education interventions to target personal control in this population, this warrants further studies in the rehabilitation setting.

With regards to the total B-IPQ mean score of 47.17 (SD: + 9.27), it is difficult to draw conclusions as the literature describes no B-IPQ cut off point as to when illness perceptions are high or low. However, the total mean B-IPQ score in our study were higher than what has previously been reported by patients with chronic obstructive pulmonary disease (49). This indicates that the JHS/EDS-HT patients in our study perceive their condition as threatening

and this may negatively influence a range of health- related outcomes, such as increased risk of disability, persistence of subjective health complaints and hence central sensitization (18, 28, 46).

A limitation of this study is the low response rate (especially in the control group, corresponding to 19.3 %), as this may make it difficult to generalize the findings. Despite the low response rate in controls the present data was congruent with other studies on SHC prevalence in the Norwegian general population (11, 12). Fortunately, there was a higher response rate for JHS/EDS-HT (44.5 %), which is not surprising, as patients often see more relevance in responding to questionnaires covering their own illness condition. In general, it should be noted that willingness to participate in epidemiological studies has declined in recent years, which means that future high participation percentages therefore may not be expected (50).

Another limitation was that the sample consisted mostly of females. This was no surprise as the disorder is seen more frequently in females (2), but naturally this limits the generalizability of our findings to men with JHS/EDS-HT.

Further, the results from the present study may be subjected to selection bias. In this study, participants with an established diagnosis were recruited by availability from two specialized hospitals. As many sufferers with JHS/EDS-HT experience the burden of misdiagnosis and wrongful treatment (9), one could assume that the present sample represents a rather resourceful selection of the real population of JHS/EDS-HT. However, this remains unknown.

Next, the use of a retrospective questionnaire for self-reported symptoms may not provide an adequate estimate of the real presence of symptoms. Even though the current questionnaire (SHC-I) is reliable, the retrospective question depends on the participant's memory, as in this case entails remembering complaints within the past 30 days.

This study has its strengths in that a reasonable number (44.5 %), of Norwegian adults with JHS/EDS-HT responded. More importantly, it provides insight into SHC and perceived illness perceptions in a Norwegian JHS/EDS-HT population, which has not previously been investigated.

In conclusion this study found that adults with JHS/EDS-HT reported significantly higher frequency and severity of subjective health complaints compared to matched controls from the general adult population in Norway. Further, the sample with JHS/EDS-HT reported low understanding of their illness and associated symptoms, and moderate beliefs that their illness could be kept under control through self-management or treatment. This may indicate one of the reasons why prognosis for these patients is poor. Our findings raise new research questions warranting better understanding of how rehabilitation health care professionals understand JHS/EDS-HT.

Clinical Messages:

JHS/EDS-HT is an under diagnosed disorder that presents clinical with widespread pain throughout the musculoskeletal system. Adults with JHS/EDS-HT also reported a higher frequency and severity of subjective health complaints compared with matched controls.

Adults with JHS/EDT-HT in this study showed maladaptive illness perceptions, characterized by perceiving the condition as difficult to understand, chronic, and only moderately controllable. Maladaptive illness perceptions are amendable, however if such perceptions remain undiscovered this could potentially complicate rehabilitation. Assessment of illness perceptions should therefore be incorporated in the multidisciplinary rehabilitation assessment.

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CONFLICT OF INTEREST

The author declares that there is no conflict of interest.

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Table I: *Demographics of the cases (JHS/EDS-HT) and controls (general population in Norway)*

Demographics	JHS/EDS-HT n=49 (100%)	Control group n=27 (100%)
Gender:		
- **Male**	3(6.1%)	2 (7.4%)
- **Female**	46 (93.9%)	25 (92.6%)
Age group:		
- **<25**	9 (18.4%)	5 (18.5%)
- **25-35**	10 (20.4%)	4 (14.8%)
- **36-45**	13 (26.5%)	5 (18.5%)
- **45-55**	13 (26.5%)	6 (22.2%)
- **>55**	4 (8.2%)	7 (26%)
BMI:		
- **<18,5**	1 (2%)	
- **18,5-24,9**	26 (54.7%)	14 (51.9%)
- **>25**	22 (43.3%)	13 (44.4%)
Education:		
- **Primary school**	10 (20.4%)	2 (7.4%)
- **High school**	21 (42.9 %)	8 (29.6%)
- **University/College (1-4 years)**	12 (24.5%)	8 (29.6%)
- **University/College (4 or more years)**	6 (12.2%)	9 (33.3%)
Employment status:		
- **Student/employed**	16 (32.7%)	17 (63%)
- **Sick leave/disability pension**	11 (22.4%)	2 (7.4%)
- **Vocational rehabilitation**	1 (2%)	3 (11.1%)
- **Employed and partially on disability pension**	10 (20.4%)	4 (14.8%)
- **Other***	9 (18.4%)	1 (3.7%)

**Different combinations of sick leave, being a student and/ or on disability pension.*

Table II: Subjective Health Complaints (SHC) covering total score, sub-scores for single items in each domain and sum score of the five domains as a measure of severity of SHC, presented as mean (SD) values for cases (JHS/EDS-HT) and controls (age and gender matched population in Norway).

	JHD/EDS-HT n=49 Mean SD	Control group n=27 Mean SD	P-value
SHC total score	32.06 ± 12.18	11.08 ± 10.07	<.001*
Headache	1.94 ± 0.25	1.50 ± 0.51	<.001*
Neck pain	1.96 ± 0.20	1.46 ± 0.51	<.001*
Upper back pain	1.71 ± 0.43	1.31 ± 0.47	.001*
Low back pain	1.98 ± 1.46	1.38 ± 0.50	<.001*
Arm pain	1.92 ± 0.28	1.40 ± 0.50	<.001*
Shoulder pain	1.92 ± 0.28	1.35 ± 0.49	<.001*
Feet pain	1.94 ± 0.25	1.31 ± 0.47	<.001*
Migraine	1.35 ± 0.81	1.23 ± 0.43	.817
Musculoskeletal complaints	14.07 ± 4.37	4.79 ± 4.69	<.001*
Palpitation	1.49 ± 0.50	1.12 ± 0.33	.001*
Heath flushes	1.57 ± 0.50	1.23 ± 0.43	.005*
Sleep problems	1.89 ± 0.32	1.38 ± 0.50	<.001*
Tiredness	1.94 ± 0.25	1.54 ± 0.51	<.001*
Dizziness	1.83 ± 0.38	1.27 ± 0.45	<.001*
Anxiety	1.34 ± 0.48	1.15 ± 0.37	.089
Depressed	1.47 ± 0.50	1.15 ± 0.37	.008*
Pseudo neurological complaints	8.63 ± 3.99	3.15 ± 3.83	<.001*
Heartburn	1.45 ± 0.50	1.12 ± 0.33	.004*
Gas discomfort	1.78 ± 0.42	1.45 ± 0.51	.006*
Diarrhea	1.40 ± 0.50	1.15 ± 0.37	.029*
Obstipation	1.43 ± 0.50	1.15 ± 0.37	.016*
Abdominal pain	1.28 ± 0.46	1.04 ± 0.20	.013*
Ulcer	1.14 ± 0.35	1.04 ± 0.20	.190
Bellyache	1.74 ± 0.44	1.08 ± 0.27	<.001*
Gastrointestinal complaints	5.68 ± 3.98	1.77 ± 2.44	<.001*
Asthma	1.25 ± 0.44	1.00 ± 0.00	.006*
Breathing difficulties	1.34 ± 0.38	1.08 ± 0.27	.013*
Allergies	1.46 ± 0.50	1.19 ± 0.40	.026*
Chest pain	1.29 ± 0.46	1.15 ± 0.39	.190
Eczema	1.33 ± 0.47	1.12 ± 0.33	.041*
Allergic complaints	1.46 ± 0.50	0.73 ± 1.46	.001*
Cough/bronchitis	1.43 ± 0.48	1.15 ± 0.37	.099
Cold/flu	1.48 ± 0.50	1.33 ± 0.48	.223
Flu/cold complaints	1.31 ± 1.57	0.70 ± 0.99	.096

*Significant difference detected between the two groups.

Table III: *Brief illness perception questionnaire items (BIPQ items), presented as mean (SD) values for cases (JHS/EDS-HT).*

BIPQ items	JHS/EDS-HT (n=47) Mean SD
Consequences[a]	6.39 ± 2.08
Timeline[a]	9.39 ± 1.52
Personal control	5.39 ± 2.3
Treatment control	4.98 ± 2.9
Identity[a]	6.7 ± 2.04
Concern[a]	4.76 ± 2.89
Understanding	3.93 ± 2.88
Emotional response[a]	5.37 ± 2.81
Total B-IPQ score	**47.17 ± 9.27**

[a] Higher score indicates more maladaptive illness perceptions

Figure I: *Flowchart; enrolment of study participants.*

Figure II*: Subjective health complaints (SHC) in cases (JHS/EDS-HT) and controls (age and gender matched population in Norway), illustrated by distribution (% in each group reporting any SHC complaint) n=49 for cases, n= 27 for controls.*

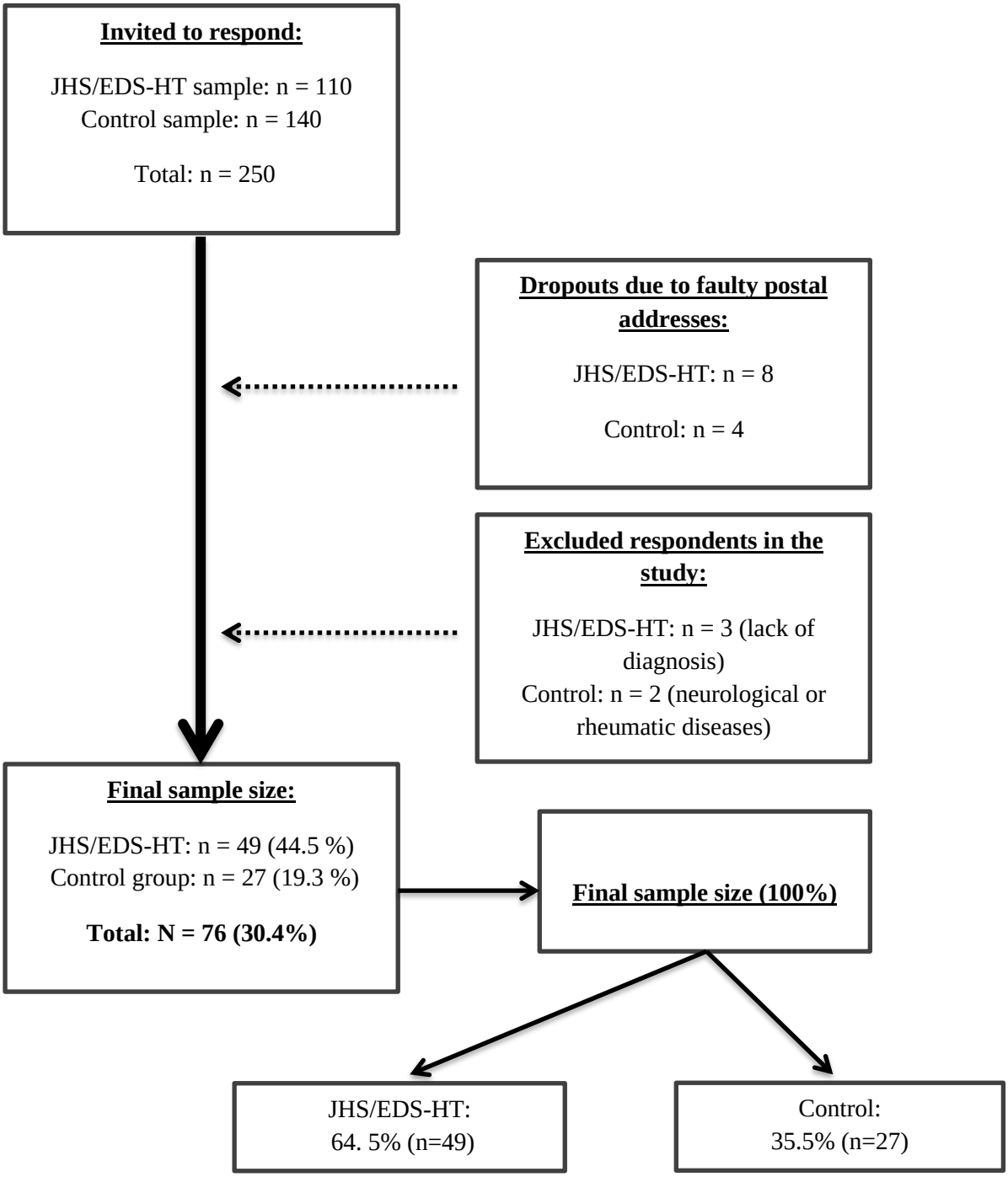

Figure I: *Flowchart; enrolment of study participants.*

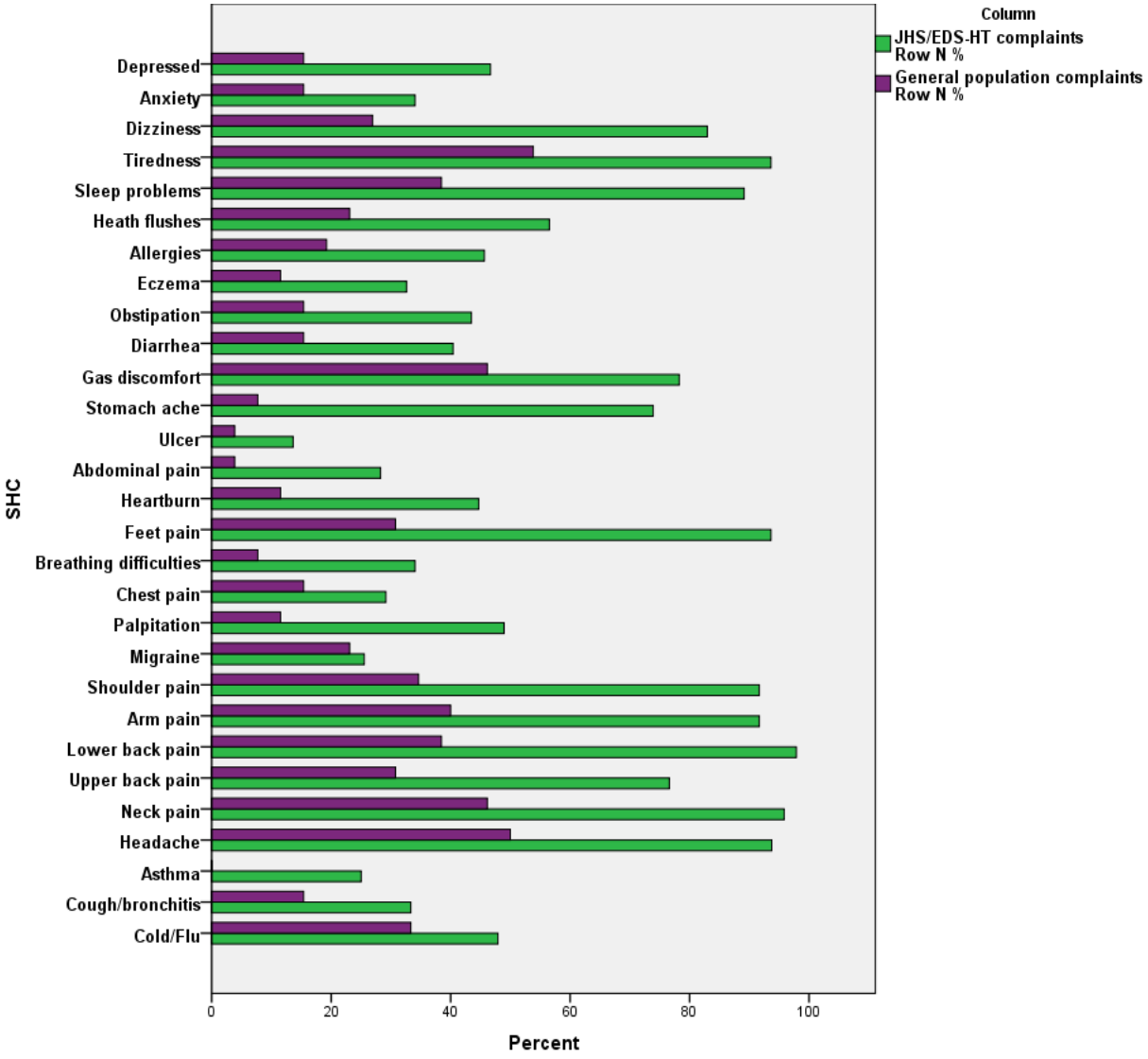

Figure II: Subjective health complaints (SHC) in cases (JHS/EDS-HT) and controls (age and gender matched population in Norway), illustrated by *distribution (% in each group reporting any SHC complaint) n=49 for cases, n= 27 for controls.*