



Development and initial validation of The Spider, a multisystem symptom impact questionnaire for patients with joint hypermobility (Part One)

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BACKGROUND

Patients with joint hypermobility report widespread chronic pain and joint dislocations as their main symptoms. However, the hEDS/HSD phenotype is variable in severity and presentation, and the consequences are known to resonate far beyond the borders of the musculoskeletal system (1). Fatigue, orthostatic intolerance, slow bowel transit, diarrhoea, constipation, and urinary incontinence are examples of 'non-musculoskeletal' symptoms that are frequently associated with generalized joint hypermobility.

When seeking diagnosis and treatment, many patients report feeling as if these symptoms are neglected at first, because they are not taken into consideration when making the diagnosis of HSD or EDS. Although the functional multisystemic symptoms do not have a central place in the diagnostic procedure, a growing body of evidence demonstrates that they significantly impact on quality of life in children and adults (2).

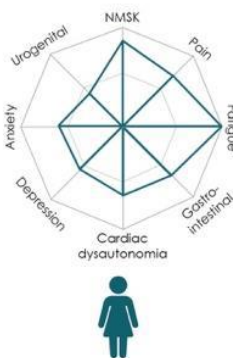
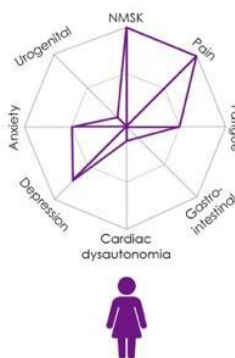
To improve insight into the symptom profile, an international research group is developing 'The Spider', a questionnaire tool which aims to evaluate the impact of important symptoms associated with HSD/hEDS. The completed spider produces a radar graph shaped as a spider web, that provides a visual overview of a patient's symptom profile (Figure 1).

In clinics, this screening tool aims to allow health care professionals to quickly identify which problems should be treated as a priority in further assessment and treatment within the multidisciplinary team. The spider may also be a useful research tool, to gain insight into the natural history of hypermobile patients and identify subgroups of patients.

Our aim is to develop The Spider into a robust screening tool and outcome measure, using the IQOLA guidelines for cross-cultural validation of health-related quality of life questionnaires (3). We report the initial stages of establishing content and face validity.



The Spider



Patients with the same diagnosis can have a very different symptom profile. The Spider graph can be used in clinics, because it provides a quick overview of the symptoms that impair daily life and require further assessment and treatment.

Evidence is emerging that patients with many non-musculoskeletal symptoms have a worse disease progression. *Examples of these multisystemic 'comorbidities' are fatigue, gastrointestinal problems, problems with blood pressure regulation, bladder problems, etc. (patient on the right).*

The Spider questionnaire can be used to detect subgroups of patients with similar symptom profiles.

Researchers are interested to find out why these patients differ from each other, how this affects functioning in daily life, and whether or not they have different genetic defects.

In addition, future research is focused on setting up more individualised treatment strategies that are better tailored to the symptom profile.



METHODS AND RESULTS

Step 1

An initial draft of The Spider consisting of 10 axes (or 'spider legs') and 60 items (questions) was created by a multidisciplinary team specialized in both musculoskeletal and non-musculoskeletal symptoms. In 2019, the questionnaire was sent to three additional experts in the field of joint hypermobility and the comorbidities, selected from the EDS Consortium.

Results and discussion

Based on the feedback, two axes were removed ('cognitive deficits' and 'allergy and mast cell activation'), because of insufficient evidence to support an association between these symptoms and EDS/HSD. A 5-part Likert scale was agreed. Furthermore, the number of

items was reduced to 25, to allow patients to fill out the questionnaire timely prior to medical consultation.

Step 2

Further face, content validity and clinical utility was evaluated by piloting The Spider in 30 adolescents and adults within the hEDS/HSD spectrum. Participants provided their opinion on the included questions, and whether or not important symptom domains were neglected.

Results and discussion

Based on their feedback, the wording in several items was adjusted (for example 'anxiety' was changed to 'worry').

CONCLUSION

Initial face and content validity of The Spider has been established. Further research into the psychometric properties such as convergent validity, known group validity and reliability are underway.

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DECLARATIONS OF INTEREST

None