



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

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BACKGROUND

Patients with joint hypermobility report widespread chronic pain, fatigue and joint instability as their main symptoms. However, the hEDS/HSD phenotype is variable in severity and presentation and extend beyond the musculoskeletal system. Fatigue, orthostatic intolerance, slow bowel transit, diarrhea, constipation, and urinary incontinence are examples of 'non-musculoskeletal' symptoms that are frequently associated with generalized joint hypermobility.

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 (1).

To improve insight into the symptom profile and to help direct and monitor patient care, 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 will allow health care professionals to quickly identify which problems should be assessed and treated as a priority within the multidisciplinary team.

The IQOLA guidelines for cross-cultural validation of health-related quality of life questionnaires is being used to produce a robust tool. Initial face and content validity has been undertaken with expert clinicians and patients.

In this abstract we report the convergent and known group validity of the pain and fatigue domains of The Spider in an adolescent population.

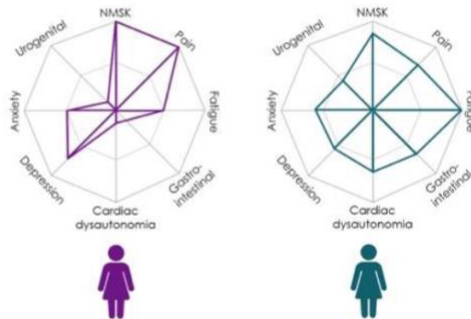


Figure 1. The Spider Spidergram: examples of different presentations across 8 symptom domains

METHODS

The study was a case control design using an online questionnaire comprised of the pain and fatigue domain questions of The Spider and the PedsQL Multidimensional Fatigue Scale (MFS) (2,3) and Cali-9.

Ethical approval was obtained from UCL Ethics committee. The questionnaire was distributed to parents of adolescents aged 13-17 and to young people aged 18 years with a diagnosis of HSD/ hEDS receiving physiotherapy treatment in a specialist hypermobility clinic and via patient charity groups (HMSA, EDS Support UK and Ehlers Danlos Society). Snowball sampling was used to expand the recruitment of adolescents with HSD/ hEDS and to recruit healthy young people for the control group. Data were collected using REDcap software and statistical analysis performed in SPSSv26. Pearson correlations were used to test the convergent validity and independent T-tests were used to examine known-group validity.

RESULTS

216 fully completed questionnaires were analysed. 177 participants were diagnosed with HSD/ hEDS, and 39 were healthy control participants.

A strong negative correlation between the fatigue domain of The Spider and the MFS was shown ($r = -0.7$, $p = 0.01$) (Figure 2).

A strong positive correlation between the pain domain of The Spider with the Cali-9 was also shown ($r = 0.74$, $p = 0.01$) (Figure 3).

A significant difference ($p < 0.001$) between healthy control adolescents and those with HSD/hEDS was found for both the pain and fatigue domain scores of The Spider indicating that the pain and fatigue scales can discriminate between symptomatic and asymptomatic young people.

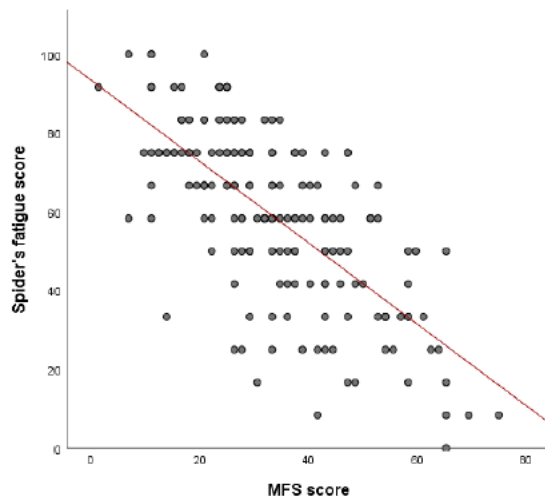


Figure 2: Scatterplot of The Spider's fatigue score with MFS score ($r=-0.70$)

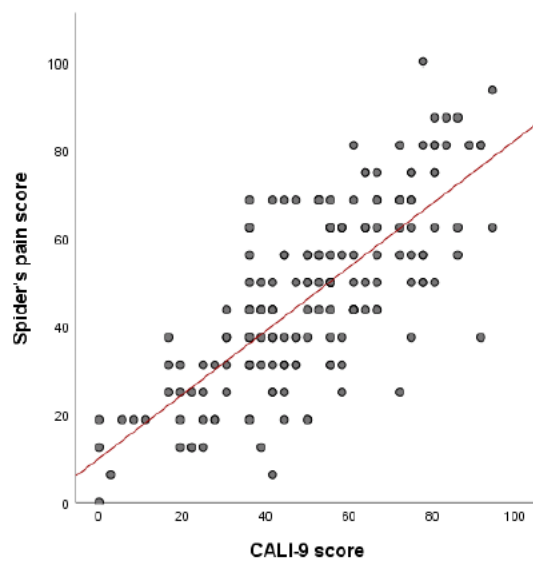


Figure 3: Scatterplot of The Spider's pain score with the Cali-9 ($r=0.74$)

CONCLUSION

These results show strong convergent and known group validity for the fatigue and pain domains of The Spider in adolescent hSD/hEDS populations.

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DECLARATIONS OF INTEREST

None