A SIMPLE QUESTIONNAIRE TO DETECT HYPERMOBILITY: AN ADJUNCT TO THE ASSESSMENT OF PATIENTS WITH DIFFUSE MUSCULOSKELETAL PAIN

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SUMMARY The aim of the study was to develop a simple and reproducible self-reporting questionnaire that identifies individuals with hypermobility. Two hundred and twelve consecutive hypermobile female new attendees to the hypermobility clinic at two London teaching hospitals and a random selection of 57 healthy volunteers completed a 10-part questionnaire. Questions were selected from clinical experience (RG), and assessed musculoskeletal symptoms and past and present physical agility. Of the 212 cases, 30 were hypermobile with no other underlying disorder and 182 fulfilled the 1998 Brighton criteria for benign joint hypermobility syndrome (BJHS). Odds ratios for the presence of hypermobility were calculated for each question. Six questions were found to be significant and the model of 'best fit' for sensitivity and specificity contained five of these. To demonstrate the reproducibility of the five-part questionnaire a second cohort of 170 hypermobile cases with BJHS and 50 controls was surveyed. Analysis demonstrated that a positive answer to any two questions in the five-part questionnaire gave the highest combined sensitivity and specificity for detecting hypermobility. The sensitivity and specificity was 84% and 89% respectively in the first cohort and reproduced with values of 84% and 80% in the second cohort. Overall the questionnaire correctly identified 84% of all cases and controls. This simple and reproducible questionnaire for detecting hypermobility could be of particular use as an adjunct in the clinical assessment of chronic, diffuse pain syndromes where hypermobility is often missed yet is potentially treatable. (*Int J Clin Pract* 2003; **57(3)**: 163-166)

Patients with focal or diffuse chronic musculoskeletal pain for which no identifiable degenerative or inflammatory cause is found should be assessed for hypermobility syndrome. Hypermobility, an inherent laxity of ligaments, may be present in up to 10% of individuals in western populations[1,2] and is more common in females than males.[3] The true prevalence is unclear as there have been few large population studies.

In the absence of a heritable disorder of connective tissue (HDCT) such as Marfan and Ehlers-Danlos syndrome,[4,5] hypermobility was once considered simply to represent the upper end of a Gaussian distribution for normal joint range of movement.[6] It is often asymptomatic and may even be advantageous in, for example, dancers and musicians. The association, however, between hypermobility, chronic pain and distress is now well documented,[7] and was first described 35 years ago by Kirk *et al*.[8]

The commonest condition seen is termed the benign joint hypermobility syndrome (BJHS), considered by many a *forme fruste* of an HDCT and associated with a number of clinical features that constitute internationally recognised diagnostic criteria.[9] Despite this, many patients with hypermobility and chronic pain remain undiagnosed. For many, their symptoms could be treated effectively with a combination of physiotherapy and, where appropriate, pain management.[10]

In practice, the diagnosis of hypermobility requires a physical examination and the application of either the Beighton[11] or Contompasis[12] score. While these scoring systems are important for epidemiological studies, they are not suitable for self-assessment questionnaires. In this study we present a simple five-part self-reporting questionnaire that identifies past or present hypermobility, and discuss its place in the clinical setting.

METHOD

Females aged between 16 and 80 years presenting to the rheumatology outpatient clinic at two London teaching hospitals were asked to complete a 10-part questionnaire structured in a 'yes/no' format. The clinics constituted both general rheumatology patients and cases referred for symptoms related to but as yet not diagnosed as a hypermobility syndrome. Nursing, physiotherapy and hospital administrative staff also took part in the study. Local hospital ethical committee approval was sought and granted.

Data were collected on age and ethnicity, and a history and physical examination were undertaken by a clinician (AH or RG). The nine-point Beighton criteria for hypermobility[11]

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Table 1. The nine-point Beighton hypermobility score[11]

	Right	Left
Ability to:		
1. Passively dorsiflex the fifth metacarpo-phalangeal joint to 90 degrees	1	1
2. Oppose the thumb to the volar aspect of the ipsilateral forearm	1	1
3. Hyperextend the elbow to ≥10 degrees	1	1
4. Hyperextend the knees to ≥10 degrees	1	1
5. Place the hands flat on the floor without bending the knees	1	
Maximum total	9	

One point is gained for each side for manoeuvres 1-4

Table 2. Revised diagnostic criteria for BJHS[9]

Major criteria
1. A Beighton score of 4/9 or more (currently or historically)
2. Arthralgia for longer than three months in four or more joints

Minor criteria
1. A Beighton score 1, 2 or 3/9 (0 if aged 50+ years)
2. More than three months arthralgia in 1-3 joints or back pain, spondylosis, spondylolysis
3. Dislocation/subluxation in more than one joint, or in one joint on more than one occasion
4. Three or more soft tissue rheumatic lesions
5. Marfanoid habitus
6. Abnormal skin: striae, hyperextensibility, thin skin, papyraceous scarring
7. Eye signs: drooping eyelids, myopia, or antimonogoloid slant
8. Varicose veins, hernia, or uterine/rectal prolapse

BJHS is diagnosed in the presence of two major criteria, one major and two minor criteria, or four minor criteria; two minor criteria suffice where there is an unequivocally affected first-degree relative.
BJHS is excluded by the presence of Marfan or Ehlers-Danlos syndromes (other than the EDS hypermobility type (formerly EDS III). Criteria major 1 and minor 1 are mutually exclusive, as are major 2 and minor 2.

(Table 1) and the revised Brighton 1998 criteria for BJHS[9] (Table 2) were used to classify individuals clinically as hypermobile with or without BJHS. Patients with an HDCT were not included in the survey.

Individuals answered the following questions:

1. Can you now (or could you ever) place your hands flat on the floor without bending your knees?
2. Can you now (or could you ever) bend your thumb to touch your forearm?
3. Do most of your joints click all the time?
4. As a child were you said to be suffering from 'growing pains'?
5. As a child did you amuse your friends by contorting your body into strange shapes *or* could you do the splits?
6. As a child or teenager did you practise gymnastics or ballet dancing?
7. As a child or teenager did your shoulder or kneecap dislocate on more than one occasion?
8. Do you consider yourself double-jointed?
9. Is any other member of your family double-jointed?
10. If you have ever had a local anaesthetic, did you think that it was as effective as it should have been?

The questions were selected on the basis that they were often asked within local clinical practice and described common rheumatic complaints. The expectation was of an answer 'yes' to any of questions 1-9 in hypermobile individuals. Question 10 was based on the finding that individuals with BJHS may have a resistance to the effect of local anaesthetic.[13,14]

Table 3. Five-part questionnaire for identifying hypermobility

1. Can you now (or could you ever) place your hands flat on the floor without bending your knees?
2. Can you now (or could you ever) bend your thumb to touch your forearm?
3. As a child did you amuse your friends by contorting your body into strange shapes or could you do the splits?
4. As a child or teenager did your shoulder or kneecap dislocate on more than one occasion?
5. Do you consider yourself double-jointed?

The data were analysed by case control methods using STATA,[15] examining each question individually for an association with hypermobility with or without other features of BJHS. Odds ratios were derived by logistic regression (95% CI), taking account of age. Questions that were significant independent of the presence or absence of BJHS were explored further to identify combinations of answers that gave the greatest combined sensitivity and specificity for hypermobility. A five-part questionnaire was identified that could correctly identify 84% of all individuals (Table 3).

Any remaining questions found to be significant for BJHS *per se* were also analysed to see if the questionnaire could identify BJHS rather than hypermobility. Although odds ratios for these questions were significant for BJHS, the specificity of each question was poor. As such they were considered unsuitable as potential screening questions.

In the second and final stage of the study a different cohort was surveyed by the same method to assess the reproducibility of the five-part questionnaire for hypermobility.

RESULTS

Two hundred and twelve cases with hypermobility and 59 controls took part in the first stage of the study. Of the 212 cases, 30 were hypermobile with no other underlying disorder and 182 fulfilled the Brighton 1998 criteria for BJHS. The second cohort consisted of 170 cases, all with BJHS, and 50 controls. More than 95% of cases and controls were caucasian and, as such, differences in ethnicity were not assessed. The response rate for each question in both surveys ranged between 93% and 100%, and although cases were on average 10 years older than controls in both surveys, age was not found to have a confounding effect on any of the questions. The means and range for age in cases and controls were similar between the first and second cohorts.

The distribution of responses and the odds ratios for each question are shown in Table 4. Questions 1, 2, 5 and 6 were identified as having significant odds ratios and a specificity and sensitivity for hypermobility greater than 60%. All controls answered questions 7 and 8 as 'no'. Any associations between hypermobility and a positive answer to questions 3, 4, 9 and 10 were not significant.

Analysis demonstrated that a positive answer to any two of five questions (1, 2, 5, 7 and 8) gave the greatest overall sensitivity and specificity for hypermobility (Table 5). The

Table 4. Distribution of answers to each question and the odds ratios for hypermobility

Question number	Hypermobile with BJHS (A)		Hypermobile without BJHS (B)		Non-hypermobile (C)		Odds ratios for hypermobility with BJHS (A *vs* C)	Odds ratios for hypermobility without BJHS (B *vs* C)
	Yes	No	Yes	No	Yes	No		
Cohort 1								
1	150	31	23	7	19	38	10.60 (5.32-21.11)	4.62 (1.71-12.49)
2	125	55	21	9	3	54	43.06 (12.87-144.06)	30.6 (7.54-124.17)
3	105	75	5	25	11	46	6.01 (2.91-12.39)	0.91 (0.28-2.96) ns
4	92	86	8	22	10	47	5.15 (2.44-10.87)	2.07 (0.68-6.53) ns
5	112	66	15	15	7	50	13.21 (5.64-30.95)	6.57 (2.17-19.86)
6	118	64	12	18	19	38	5.00 (2.61-9.58)	2.89 (1.09-7.61)
7	36	145	2	28	0	55	-	-
8	90	88	6	24	0	57	-	-
9	83	93	2	26	11	44	3.77 (1.82-7.79)	0.37 (0.07-1.85) ns
10	73	99	4	24	11	42	2.85 (1.37-5.93)	0.53 (0.13-2.14) ns
Cohort 2								
1	126	39			16	34	6.87 (3.42-13.47)	
2	122	42			15	35	6.78 (3.37-13.64)	
5	109	56			5	45	17.52 (6.58-46.60)	
7	62	102			0	50	-	
8	109	53			0	49	-	

ns, not significant

Table 5. Comparison of four models for the shortened questionnaire: data from cohort 1

Model	Odds ratio* for hypermobility (95% CI)	Sensitivity %	Specificity %	Correct classification %	LR Chi Sq
Answer 'Yes' to any one of Q1,2,5,6,7 or 8	20.31 (8.34-49.46)	96	47	85	53.4
Answer 'Yes' to any one of Q1,2,5,7 or 8	22.92 (10.12-51.96)	94	58	86	67.8
Answer 'Yes' to any pair of Q1,2,5,6,7 or 8	23.68 (11.00-50.96)	89	74	86	80.3
Answer 'Yes' to any pair of Q1,2,5,7 or 8	37.26 (14.72-94.35)	83	89	84	95.1

*odds ratio calculated from group (A + B) vs group C (see Table 4)

addition of any further questions had a significant negative effect on the specificity. Notably, question 6 did not add to the cases of hypermobility already identified by the other five questions but lowered the specificity by the inclusion of extra controls.

The data for sensitivity and specificity of the five-part questionnaire in both the first and second cohorts is shown in Table 5. The five-part questionnaire correctly identified 84% of individuals with a sensitivity of 77-85% and specificity of 89% in the first cohort and 85% and 80% respectively in the second cohort. The difference in sensitivity between cases with BJHS and those without in the first cohort was small, suggesting the questions identified hypermobility rather than any other particular feature of BJHS, in keeping with the odds ratio data (Table 6).

DISCUSSION

We have developed a reproducible and simple self-reporting five-part questionnaire as a means of identifying individuals with hypermobility. We anticipate that this questionnaire will be of greatest value in the clinic setting when assessing patients with focal or diffuse chronic musculoskeletal pains in whom no clear evidence for degenerative or inflammatory arthropathy has been found.

The most frequently encountered hypermobility syndrome is BJHS,[10] considered synonymous with the hypermobility type of Ehlers-Danlos syndrome (formerly Ehlers-Danlos type III).[16] The multisystemic nature of this condition is well documented and many consider it a connective tissue disorder in its own right, manifesting several features that overlap with rarer disorders of hypermobility such as Marfan syndrome, 'classical' Ehlers-Danlos (formerly types I and II), and osteogenesis imperfecta.[10,16] Inappropriately, the problems associated with hypermobility are often thought to diminish with time as the degree of hypermobility wanes with age. Likewise, it is often assumed the problem is purely an articular one, and one for which there is no satisfactory treatment. The result is a wide diversity of opinion and practice, with many patients remaining undiagnosed and their symptoms sometimes trivialised.[7,15] Though often difficult, the symptoms of BJHS can, however, be treated effectively with a combination of appropriate physiotherapy and pain management.[10,19] Many patients with BJHS also report a range of extra-articular symptoms, some of which are related to psychological stresses such as anxiety and depression, but others possibly associated with autonomic dysfunction.[20] This latter finding is an area of current research and may explain why it is that some subgroups of hypermobile

Table 6. Case control data for answering 'yes' to any two items in the five-part questionnaire

			Answer 'yes' to any two questions		Five-part questionnaire		
Group	Mean age years (sd)	No. in survey	Yes	No	Sensitivity	Specificity	Correctly classified
Cohort 1							
Cases (with BJHS)	36 (12)	182	153	29	84%	89%	85%
Controls	43 (14)	57	6	51			
Cohort 1							
Cases (without BJHS)	34 (11)	30	23	6	77%	89%	85%
Controls	43 (14)	57	7	51			
Cohort 2							
Cases	33(11)	170	143	27	84%	80%	83%
Controls	42 (12)	50	10	40			

patients appear more prone to multiple extra-articular symptoms than others. Whatever the underlying mechanisms of pathology may turn out to be, identifying the presence of hypermobility is clearly an important early step in the diagnosis.

Hypermobility remains poorly explored in the general population. Large-scale studies have been hampered by the lack of a validated, self-reporting screening questionnaire. A further application of our questionnaire could therefore be its use as a screening tool in epidemiological surveys. Although the Beighton and Contompasis scores are invaluable clinical tools for identifying and quantifying hypermobility, they do not lend themselves readily to self-assessment and do not take account of hypermobility at sites other than the fingers and thumb, elbow, lower back and knees, albeit these sites are the ones most often affected. There is now evidence to suggest that pauciarticular (1-3 joints) hypermobility is more prevalent than polyarticular hypermobility in the general population.[13,21] The use of a 'cut-off' score of 4/9 on the Beighton scale, for example, is likely to underestimate the true prevalence of hypermobility syndromes. It should be noted that this is taken into account in the 1998 Brighton diagnostic criteria for BJHS[9] by inclusion of pauciarticular hypermobility as one of the minor criteria. It is also the case that while two of the items in our five-part questionnaire ask about hypermobility at specific joints, the other three items do not and focus on historical and general aspects of agility. The value of such a simple self-reporting screening questionnaire is self-evident, and further population studies with our questionnaire are now required.

CONCLUSION

Hypermobility should be considered in the differential diagnosis of patients presenting with chronic pain and recurrent soft tissue lesions. As an aide-memoire for the clinician we have developed both a sensitive and specific simple questionnaire to identify hypermobility. Further studies may show this questionnaire to be of value as a screening tool in population surveys.

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