The relationship between benign joint hypermobility syndrome and psychological distress: a systematic review and meta-analysis

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Abstract

Objective. This study examines the reported evidence of an association between benign joint hypermobility syndrome (BJHS) and psychological symptoms.

Methods. A systematic review of published (AMED, CINAHL, MEDLINE, EMBASE, PubMed, Cochrane Library) and unpublished research databases (OpenGrey, the World Health Organization (WHO) International Clinical Trials Registry Platform, Current Controlled Trials, the UK National Research Register Archive) was performed from their inception to January 2013. Studies assessing the prevalence and incidence of psychological conditions for people diagnosed with BJHS were included. Meta-analysis assessing the odds ratio (OR) and standardized mean difference in severity of psychological conditions was performed. Methodological quality was assessed using the Critical Appraisal Skills Programme (CASP) appraisal tools.

Results. Fourteen papers including 3957 participants, 1006 people with and 2951 controls without BJHS were eligible. The overall methodological quality was moderate. The results indicated that people with BJHS experience significantly greater perceptions of fear and more intense fear ($P < 0.05$) and have a higher probability of demonstrating agoraphobia ($P < 0.05$), anxiety (OR 4.39, 95% CI 1.92, 10.40), depression (OR 4.10, 95% CI 1.79, 9.41) and panic disorders (OR 6.72, 95% CI 2.22, 20.35) than those without BJHS ($P < 0.005$). Neither anxiety nor depression have been assessed in childhood populations.

Conclusion. People with BJHS commonly exhibit a range of symptoms related to anxiety and depression. Considerable emotional symptoms accompany BJHS. Further study is warranted to explore how these results relate to non-Mediterranean populations and children. However, the data suggest that targeting psychological symptoms could be an important approach to managing the range of symptoms reported in these patients.

Key words: psychological entity, benign joint hypermobility syndrome, panic disorder, mental health, assessment.

Introduction

Benign joint hypermobility syndrome (BJHS) is a heritable connective tissue disorder that is characterized by excessive joint flexibility and musculoskeletal dysfunction [1, 2]. While joint hypermobility may be seen throughout the population, and can be measured with the Beighton scale [3], BJHS is classified as excessive joint mobility with symptoms such as pain. The Brighton classification was developed to incorporate both joint flexibility and pain, and this scale is now widely regarded as the accepted means of clinically diagnosing this clinical population [4]. BJHS is more commonly seen in females compared with males and in children and adolescents [5]. While the prevalence of hypermobility in children is high, only a proportion of hypermobile children complain of pain [6].
People with BJHS may present with traumatic OA, subluxation or dislocation of peripheral joints, tendinopathy or bursitis. Previous authors have acknowledged that such conditions can lead to persistent or chronic pain states [7, 8]. The association of chronic pain and psychological distress, principally as anxiety or depression, is well documented in the literature for other populations such as people with OA, RA and lupus [9, 10]. Previous authors have also acknowledged that psychological factors can potentially modify the pain experienced by patients [11, 12]. However, any association of these mental health problems and BJHS has been less clearly defined, with no previous research synthesis undertaken to evaluate the entire evidence base on this pathology. Therefore the purpose of this study is to address this limitation and to examine the available literature to determine whether there is a relationship between BJHS and psychological distress or psychological pathologies.

Materials and methods

Types of studies

Case–control and cohort study designs were included to assess the prevalence and incidence of psychological distress for people diagnosed with BJHS. Single-case studies were excluded. No restriction was placed on the language of the article or date of publication.

Participants

Cases included in the review were those with a clinical diagnosis of BJHS. This was defined as symptomatic (pain) joint hypermobility with a Beighton score ≥ 4 [4]. No restriction was placed on the place of recruitment. Participants who were diagnosed with other joint laxity/connective tissue disorders such as Marfans and Ehlers–Danlos were excluded, with the exception of Ehlers–Danlos type III. People awaiting surgical interventions for musculoskeletal pathology were excluded since this was regarded as a potential source of anxiety or depression that may not be similar to those not awaiting surgery.

Outcome measurement

The primary outcome measurement was the prevalence of anxiety in those people with BJHS compared with those without. Secondary outcome measurements included the prevalence of fear, panic, phobia, depression and dysthymic disorders between these two populations.

Search strategy

The search strategy was developed based on the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) guidelines [13]. The search was therefore divided into three domains: anatomical, pathological and outcome based. Search terms relating to peripheral and central joint regions, hypermobility and joint flexibility to identify those with BJHS were used in addition to terms to describe anxiety, depression and psychological pathologies. These are presented in supplementary Table S1, available at Rheumatology Online as a MEDLINE search, which was adapted for the other databases.

The electronic databases—AMED, CINAHL, EMBASE, psycINFO, MEDLINE and the Cochrane Library—were searched from their inception to January 2013. Unpublished electronic databases such as OpenGrey, the World Health Organization (WHO) International Clinical Trials Registry Platform, Current Controlled Trials and the UK National Research Register Archive were also reviewed from their inception to January 2013. The reference lists of each eligible article were scrutinized for any additional articles. Finally, the corresponding authors from all included articles were consulted in order to identify any additional articles and, where necessary, were asked for clarification on their data.

Study eligibility

Based on the eligibility criteria above, two reviewers (T.S., V.E.) independently reviewed the titles and abstracts from potentially relevant articles identified through the search strategy. The full texts of all potentially eligible articles were reviewed before making a final decision on eligibility. Studies were excluded if they did not address the research question.

Data collection

Data were entered onto a pre-defined data extraction table. Data extracted included characteristics of the participants (both BJHS and asymptomatic controls) including age, gender, method of diagnosis, degree of joint hypermobility (frequently assessed using the Beighton scoring system), co-morbidities, method of assessing psychological status and subsequent findings. This was performed by one reviewer (V.E.) and was verified by a second (H.B.). Any disagreements in data extraction were resolved through discussion between the reviewers.

Critical appraisal

Each included article was critically appraised using the Critical Appraisal Skills Programme (CASP) Case Control appraisal tool [14] or the CASP Cohort Study tool [15], depending on the study design. These tools have been widely adopted for the review of previous musculoskeletal clinical studies [16–18]. Each included article was reviewed by one reviewer (V.E.) and independently verified by a second (H.B.). Any disagreements in appraisal score were discussed and resolved through a third reviewer (T.S.). This systematic review was registered with PROSPERO, an international database of systematic reviews in health and social care (http://www.crd.york.ac.uk/NIHR_PROSPERO, registration number CRD42012002372).

Data analysis

Study methodological heterogeneity was assessed visually using the data extraction table. If methodological heterogeneity was evident, a qualitative narrative review of results was conducted. If methodological homogeneity was evident in participant characteristics, follow-up
period and data collection methods, a meta-analysis was deemed appropriate. Statistical heterogeneity was evaluated through observation of forest plots and by using the chi-squared and $I^2$ statistical tests. When $P > 0.10$ and $I^2 \geq 20\%$, a random-effects model was undertaken. When $P < 0.10$ and $I^2 < 20\%$ a fixed-effects model was employed. For continuous outcomes, mean differences or standardized mean differences (SMDs) were calculated. For dichotomous outcome measurements, odds ratios (ORs) were calculated. For each statistic, a 95% CI and $P$-value was presented.

The assessment of potential small-study publication bias was made by evaluating the asymmetry of a funnel plot for the primary outcome (anxiety). No specific exclusions were placed on analysing childhood cohorts, although a specific subgroup analysis of psychopathology in the childhood cohort was undertaken.

All statistical analyses were conducted on RevMan version 5.1 (Nordic Cochrane Centre, Cochrane Collaboration, Copenhagen, Denmark, 2011). No ethical approval was required for this study.

Results

Search strategy results

A summary of the results of the search strategy is presented in the PRISMA flow chart shown in Fig. 1. This indicates that while 172 articles were identified, a total of 14 articles satisfied the eligibility criteria and were included in the review.

Methodological appraisal

The results of the critical appraisal process are summarized in supplementary Tables S2 and S3 (available at Rheumatology Online). Overall, this indicated that the current evidence base displayed moderate methodological quality.

Seven articles were case–control study designs and thus were analysed using a case–control CASP tool (supplementary Table S2, available at Rheumatology Online). The strengths of the literature included the recruitment of clinically representative participants, permitting the generalizability of the study findings to clinical populations in all studies. Furthermore, all studies related their findings to the previous literature. The outcomes were assessed using reliable and valid measurements for this population and were clearly analysed using inferential statistical tests. However, two studies did not clearly define the origin or recruitment processes adopted for their samples [19, 20] and two studies did not match the case to the control cohorts [21, 22]. Blinding assessors to the allocation of cases or controls was clearly demonstrated by all but two studies [19, 20]. Only one study based their sample size on a power calculation [23].

Seven studies used a cohort study design (supplementary Table S3, available at Rheumatology Online). These were analysed using the CASP cohort appraisal tool. The strengths of the current evidence base included a clearly defined research question and clarity in recruiting the study cohorts. The results were also clearly related to the previous literature and were generalizable to the current clinical population. Similar to the case–control studies, weaknesses within the literature included poor analysis of findings using CI data, which was only presented in the article by Bulbena et al. [24]. Assessor blinding and minimizing bias for data collection were only clearly defined in Bulbena et al. [24–26] and Ruperto et al. [27]. Two studies demonstrated particularly low methodological quality [27, 28], where minimal follow-up periods, limited control of confounding variables and limited detail on recruitment processes were provided.

Assessment of small-sample publication bias

The funnel plot to assess small-sample publication bias demonstrated a broadly symmetrical shape for the assessment of the anxiety score, the primary outcome measurement (supplementary Fig. S1, available at Rheumatology Online). Therefore it is possible to conclude minimal risk of small-sample publication bias for this evidence base.

Characteristics of included studies

A summary of the study characteristics for the 14 included studies is presented as Table 1. A total of 3957 participants were included in the analyses from the 14 studies. This included 1006 people with symptomatic BJHS (Beighton score $\geq 4$) and 2951 controls without joint hypermobility and symptoms (Beighton score $<4$). The cohort consisted of 1507 males and 2066 females. The proportion of males/females was not presented in three studies [20, 21, 28]. The mean age was presented in all but the article by Gratacos et al. [20]. This indicated a mean age of 29.12 years (S.D. = 12.0), ranging from 11.9 [28] to 48.1 years [21]. The specific age ranges, when documented, are presented in Table 1. Only Fatoye et al. [28], Ruperto et al. [27] and Pailhez et al. [29] recruited solely childhood populations. Three studies recruited their participants from school settings [27–29] and one study from a university [30], three studies were based in primary care/community health care settings [23–25] and seven studies were based in hospital outpatient departments [1–22, 26, 31, 32]. Studies were largely conducted in Mediterranean countries. Eight studies originated from Spain, two from Italy, one from France, one from Turkey and one from Israel, while the study by Fatoye et al. [28] derived from Scotland. All studies adopted a diagnosis of BJHS based on a Beighton score $\geq 4$ and musculoskeletal pain, with the exception of the study by Bulbena et al. [26], which used the Hospital del Mar criteria. Six articles provided mean Beighton scores ranging from 5.4 [31] to 7.2 [21].

Clinical results

The results of the meta-analyses are presented in Table 2.
Primary outcome measure

Anxiety

The results of the meta-analysis from three studies [21, 31, 32] indicated a four times greater probability of anxiety in those with BJHS compared with controls (OR 4.39, 95% CI 1.92, 10.4, \( P = 0.005 \); Fig. 2).

Seven studies assessed the difference in anxiety scores between those with BJHS and those without [19, 21, 24, 26, 30–32]. The results from the meta-analysis indicated a statistically significant difference between the groups, with greater severity of anxiety symptoms in those with BJHS compared with the control group (SMD = 0.53, 95% CI 0.31, 0.74, \( P < 0.001 \); Fig. 3).

Two studies specifically assessed social anxiety using the social anxiety score [24, 30]. This demonstrated no statistically significant difference between the groups for the total (\( P = 0.10 \)) and different subscales (\( P = 0.08, 0.16 \)) for this outcome (Table 2).

Secondary outcome measures

Depression

Three studies assessed the prevalence of depression in the BJHS population compared with non-hypermobile controls [21, 24, 32]. Overall there was a 4-fold greater probability of depression in those with BJHS compared with non-joint hypermobility controls (OR 4.10, 95% CI 1.78, 9.41, \( P < 0.001 \); supplementary Fig. S2, available at Rheumatology Online). However, when the severity of depression was assessed between these two populations, there was no statistically significant difference between the BJHS and non-BJHS cohorts (SMD = 0.32, 95% CI –0.10, 0.74, \( P = 0.14 \)).

Two studies assessed the prevalence of dysthymic disorders [21, 32]. On pooled analysis, this was not statistically significantly different between those with and without BJHS (OR 1.12, 95% CI 0.47, 2.67, \( P = 0.80 \)).

Agoraphobia

The prevalence of agoraphobia was assessed in one study only [21]. This reported a statistically significant difference between the control and BJHS groups, with a 5-fold risk of agoraphobia in those with BJHS (age- and sex-adjusted OR 5.08, 95% CI 2.06, 12.49).

Panic disorder

The prevalence of panic disorders was assessed in four studies [21, 23, 24, 32]. These indicated nearly a 7-fold greater probability of those with BJHS suffering from a panic disorder compared with non-BJHS controls (OR 6.72, 95% CI 2.22, 20.35, \( P < 0.001 \); supplementary Fig. S3, available at Rheumatology Online).

Fear

Fear was assessed in two studies [25, 29]. In both studies this was assessed using the Fear Survey Schedule to
## Table 1: Summary of cohort characteristics of the eligible studies in this review

<table>
<thead>
<tr>
<th>Study</th>
<th>Study origin</th>
<th>Sample size</th>
<th>Gender, M/F</th>
<th>Mean age (±S.D.)</th>
<th>Beighton score for cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>Baeza-Velasco et al. [30]</td>
<td>France</td>
<td>BJHS = 144</td>
<td>74/291</td>
<td>21.2 (2.11), range 18–30</td>
<td>≥5</td>
</tr>
<tr>
<td>Benjamin et al. [22]</td>
<td>Israel</td>
<td>BJHS = 13</td>
<td>61/79</td>
<td>Mean range = 23.4–39.3</td>
<td>≥5</td>
</tr>
<tr>
<td>Bulbena et al. [21]</td>
<td>Spain</td>
<td>BJHS = 114</td>
<td>N/D</td>
<td>48.1 (13.5)</td>
<td>7.2</td>
</tr>
<tr>
<td>Bulbena et al. [26]</td>
<td>Spain</td>
<td>BJHS = 77</td>
<td>323/203</td>
<td>M = 25.2 (2.9)</td>
<td>M = 3/4&lt;sup&gt;a&lt;/sup&gt;</td>
</tr>
<tr>
<td>Bulbena et al. [25]</td>
<td>Spain</td>
<td>BJHS = 182</td>
<td>597/708</td>
<td>F = 25.6 (3.2)</td>
<td>F = 4/5&lt;sup&gt;a&lt;/sup&gt;</td>
</tr>
<tr>
<td>Bulbena et al. [24]</td>
<td>Spain</td>
<td>BJHS = 29</td>
<td>73/64</td>
<td>43.0 (17.8), range 16–79</td>
<td>≥5</td>
</tr>
<tr>
<td>Ercolani et al. [19]</td>
<td>Italy</td>
<td>BJHS = 30</td>
<td>6/79</td>
<td>JHS = 32.3 (10.4)</td>
<td>≥5</td>
</tr>
<tr>
<td>Fatoye et al. [28]</td>
<td>Scotland</td>
<td>BJHS = 29</td>
<td>N/D</td>
<td>JHS = 11.9 (1.8)</td>
<td>≥6</td>
</tr>
<tr>
<td>García Campayo et al. [23]</td>
<td>Spain</td>
<td>BJHS = 59</td>
<td>38/182</td>
<td>41.3 (11)</td>
<td>≥5</td>
</tr>
<tr>
<td>Gratacós et al. [20]</td>
<td>Spain</td>
<td>BJHS = 65</td>
<td>N/D</td>
<td>N/D</td>
<td>≥5</td>
</tr>
<tr>
<td>Gurer et al. [32]</td>
<td>Turkey</td>
<td>BJHS = 40</td>
<td>9/85</td>
<td>41.1 (12.8)</td>
<td>≥5</td>
</tr>
<tr>
<td>Martín-Santos et al. [31]</td>
<td>Spain</td>
<td>BJHS = 77</td>
<td>104/136</td>
<td>38.3 (12.9)</td>
<td>≥5</td>
</tr>
<tr>
<td>Pailhez et al. [29]</td>
<td>Spain</td>
<td>BJHS = 41</td>
<td>66/84</td>
<td>16.45 (0.6), range 15–18</td>
<td>≥5</td>
</tr>
<tr>
<td>Ruperto et al. [27]</td>
<td>Italy</td>
<td>BJHS = 106</td>
<td>156/155</td>
<td>Median 10.6 (IQR 8.5–14.1)</td>
<td>≥5</td>
</tr>
</tbody>
</table>

<sup>a</sup>Hospital del Mar criteria. C: controls; M: male; F: female; IQR: interquartile range; N/D: not documented.

## Table 2: Summary of the results of the meta-analyses

<table>
<thead>
<tr>
<th>Analysis</th>
<th>Studies analysed</th>
<th>n</th>
<th>BJHS</th>
<th>Control</th>
<th>Effect estimate (95% CI)</th>
<th>P-value</th>
<th>I&lt;sup&gt;2&lt;/sup&gt;, %</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Prevalence</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
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<td></td>
</tr>
<tr>
<td>Anxiety</td>
<td>[21, 31, 32]</td>
<td>253 212</td>
<td>4.39</td>
<td>(1.92, 10.4)</td>
<td>0.005 61</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Depression</td>
<td>[21, 24, 32]</td>
<td>183 221</td>
<td>4.10</td>
<td>(1.78, 9.41)</td>
<td>0.0009 0</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Dysthymic disorder</td>
<td>[21, 32]</td>
<td>154 113</td>
<td>1.12</td>
<td>(0.47, 2.67)</td>
<td>0.80 0</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Panic disorder</td>
<td>[21–24, 32]</td>
<td>238 276</td>
<td>6.72</td>
<td>(2.22, 20.35)</td>
<td>0.0008 67</td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Severity</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Total phobia score</td>
<td>[19, 21, 31]</td>
<td>221 247</td>
<td>0.47</td>
<td>(0.08, 1.02)</td>
<td>0.09 85</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Anxiety score</td>
<td>[19, 21, 24, 26, 30–32]</td>
<td>501 948</td>
<td>0.53</td>
<td>(0.31, 0.74)</td>
<td>&lt;0.00001 63</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Depression score</td>
<td>[19, 21, 24, 30]</td>
<td>317 413</td>
<td>0.32</td>
<td>(0.10, 0.74)</td>
<td>0.14 83</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Social anxiety score (total)</td>
<td>[24, 30]</td>
<td>173 329</td>
<td>5.40</td>
<td>(1.03, 11.82)</td>
<td>0.10 57</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Social anxiety score (fear)</td>
<td>[24, 30]</td>
<td>173 329</td>
<td>1.91</td>
<td>(0.76, 4.58)</td>
<td>0.16 38</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Social anxiety score (avoidance)</td>
<td>[24, 30]</td>
<td>173 329</td>
<td>3.15</td>
<td>(0.43, 6.73)</td>
<td>0.08 54</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

<sup>a</sup>Assessed through OR. <sup>b</sup>Assessed through SMD.
assess common fears and phobias [33]. There were insufficient data to pool the results. However, in both studies participants diagnosed with BJHS demonstrated higher fear scores than those without. Pailhez et al. [29] reported a statistically significant higher fear score in those with BJHS (91.6 vs 75.9, P = 0.005), with the frequency of severe fears also significantly higher among the BJHS group (7.6 vs 11.1, P = 0.001). This was reflected in the results of Bulbena et al. [25], who also reported statistically significantly greater fear in the BJHS group (83.7 vs 66.3, P < 0.005) and severe fears (P < 0.05) compared with those with a Beighton score <4.

Total phobia score
Three studies assessed the difference in general phobia severity between those with and without BJHS [19, 21, 31]. The meta-analysis indicated no statistically significant difference between the groups with respect to this outcome (SMD = 0.47, 95% CI −0.08, 1.02, P = 0.09).

Psychopathology in childhood
Only three studies assessed psychopathology in childhood populations [27–29]. While Fatoye et al. [28] reported statistically poorer emotional functioning in their BJHS cohort (P = 0.003) using a quality of life perception tool, Ruperto et al. [27] reported no statistically significant difference between their BJHS and non-BJHS groups when assessing psychosocial summary scores from the Italian Childhood Health Questionnaire (P = 0.19) [34]. Pailhez et al. [29] assessed the intensity of fear between their BJHS and non-BJHS cohorts, reporting significantly greater fear in those with BJHS (P = 0.001). Neither anxiety nor depression has been assessed in childhood populations.

Discussion
This is the first systematic review undertaken to investigate the possible relationship between BJHS and psychological distress. The results indicate that people with BJHS have a four times greater probability of suffering from anxiety and having a significantly higher anxiety score than people without BJHS. They also have a significantly greater probability of suffering from depression and panic disorders (P < 0.05). Fear and agoraphobia also seem to be more prevalent, but have been less frequently studied, reported in only two [25, 29] and one study [21], respectively. The current evidence base is derived from adult populations, principally from the Mediterranean, which may limit the generalizability of these findings. There is a notable lack of evidence in childhood BJHS populations. The literature has not demonstrated causality, only an association between psychological pathology and BJHS. The assessment of cause and effect was not possible due to the largely cross-sectional study designs presented. Martín-Santos et al. [31] acknowledged that this will be difficult to ascertain.

This study did not aim to assess the relationship between psychological pathology and the severity of BJHS. The studies included in this review largely used the Beighton and subsequently the Brighton criteria to identify people with BJHS, and controversy remains regarding the optimal means of assessing the severity of...
symptoms [35]. Only once this has been resolved through future research will it be possible to assess the relationship between the variables. This has particular importance since Baeza-Velasco et al. [1] reported that pain was the most significant predictor of anxiety and depression in their cohort of participants with BJHS. Being able to identify whether the degree of BJHS is related to psychological distress would be a useful area for future study to identify those at greatest risk of such mental health disorders.

The results of this review indicate that people with BJHS have a significantly greater probability of psychological distress compared with those without BJHS. Numerous explanations have been hypothesized to account for this. For example, the biological link between BJHS and anxiety was highlighted by Eccles et al. [36]. They observed that people with joint hypermobility demonstrate similar autonomic cardiovascular abnormalities to those with postural tachycardia syndrome [36]. Therefore the person may experience abnormal reactive autonomic nervous system, particularly increases in heart rate on standing, which may manifest as an anxiety symptom [36, 37]. Second, Gurer et al. [32] attributed anxiety symptoms in people with BJHS to a greater perception of joint instability and frequency impacting on activities of daily living, but without understandable antecedence. Therefore understanding the condition may be a key feature in this cohort’s psychological presentation. Further study to investigate the relationship between these features may be beneficial to increase our understanding of this condition.

Since the review concluded that psychopathology is clearly associated with BJHS, management of the condition should include ensuring anxiety, depression, panic and other pathologies are identified and addressed where present. Relaxation training and active exercises are currently prescribed to people with BJHS [38, 39]. These interventions also demonstrate clinical efficacy in allaying stress, panic, anxiety and depression symptoms in other populations [38, 39]. Non-pharmacological treatments are considered the cornerstone treatment for people with BJHS [38]. Nonetheless, no studies have assessed the effects of such interventions on psychological outcomes. Further study of such outcomes is therefore warranted as a priority for this population.

The studies included in this review were based on cohorts from across the world. Bulbena et al. [24] acknowledged that anxiety, as well as panic and depression, may be expressed differently in certain contexts through cross-cultural differences. Previous studies of musculoskeletal populations have acknowledged cross-cultural differences in pain perception and statistically significant differences in the coping strategies questionnaire responses on diverting attention, catastrophizing, ignoring sensations, praying/hoping, guarding and resting [40–42]. Ferreira-Valente et al. [42] reported differences in the risk of depression and distress in those from Portugal compared with North America. The assessment of psychological factors for those with BJHS should be further explored in other cultures and geographical regions, including Northern Europe, North America, Asia and Australasia.

Conclusion

People with BJHS have a four times greater probability of suffering from anxiety and have a significantly higher anxiety score than people without hypermobility. They also have a greater probability of suffering from depression and panic disorders. Fear and agoraphobia also seem to be more prevalent, but have been less frequently studied. The current evidence base is derived from adult, principally Mediterranean, populations, which may limit the generalizability of these findings. There is a notable lack of evidence for child populations.

Further research is required to better understand the potential importance of psychological morbidity on the assessment and management of children and adults with BJHS. A second research priority is to assess possible interventions that may assist in managing such psychological disorders in this population.

Rheumatology key messages

- BJHS patients commonly exhibit a range of symptoms related to anxiety and depression.
- BJHS patients experience significantly greater perceptions of fear, agoraphobia and panic disorders

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Supplementary data

Supplementary data are available at Rheumatology Online.

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