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The views of people with Joint Hypermobility Syndrome on its impact, management and the use of patient-reported outcome measures. A thematic analysis of open-ended questionnaire responses.

SHORT RUNNING TITLE

Joint Hypermobility Syndrome: patient views.

AUTHORS AND AFFILIATIONS

Shea Palmer^a, Keely Bridgeman^a, Isabelle Di Pierro^a, Rosanna Jones^a, Caitlin Phillips^a, Megan Wilson^a

^aDepartment of Allied Health Professions, Faculty of Health & Applied Sciences, University of the West of England, Bristol, BS16 1DD, United Kingdom

CORRESPONDING AUTHOR

Professor Shea Palmer, Department of Allied Health Professions, Faculty of Health & Applied Sciences, University of the West of England, Blackberry Hill, Bristol, UK, BS10 1DD. Tel: +44 (0)117 3288919. Email: Shea.Palmer@uwe.ac.uk.

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CONFLICT OF INTEREST STATEMENT

There are no conflicts of interest for any of the authors.

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The views of people with Joint Hypermobility Syndrome on its impact, management and the use of patient-reported outcome measures. A thematic analysis of open-ended questionnaire responses.

ABSTRACT

Background: Joint Hypermobility Syndrome (JHS) has been reported to have widespread impacts on people with the condition. However, our understanding of those impacts is still developing and we do not know if they can be captured effectively using patient-reported outcome measures.

Aims: To explore written qualitative comments from previously administered questionnaires to identify the impacts of JHS and any issues related to using patient-reported outcome measures to assess those impacts.

Method: Previous research administered a draft condition-specific questionnaire and Short Form-36 questionnaire to adult members of a patient organisation in the United Kingdom, incorporating an open text box for further comments. Those comments were transcribed, anonymised and analysed using thematic analysis. A coding list, themes and sub-themes were developed through double coding, parallel independent analysis and consensus.

Results: 393 of 614 eligible questionnaires (64%) contained qualitative comments and were analysed (mean respondent age 41 years, mean Bristol Impact of Hypermobility questionnaire score 228/360, 95% women). Three main themes were identified: 1) 'Impacts of living with JHS', 2) 'Management strategies for JHS' and 3) 'Measurement and research into JHS'. Participants highlighted a range of impacts of

JHS, incorporating physical, social and psychological domains. Respondents described difficult journeys to diagnosis, and feeling unsupported and misunderstood by their peers and healthcare professionals. They detailed helpful strategies for managing their condition and provided useful comments on using questionnaires to assess JHS.

Conclusions: The study yielded valuable findings that can be used to directly inform the assessment and management of JHS.

KEY WORDS

Hypermobility, joint; Patient reported outcome measures; Qualitative research; Surveys and questionnaires.

MAIN TEXT

INTRODUCTION

Joint Hypermobility Syndrome (JHS) is a heritable connective tissue disorder associated with several adverse impacts (Ross and Grahame, 2011), including pain, impaired proprioception, soft tissue injuries and joint dislocations, premature osteoarthritis and autonomic dysfunction. It is linked with other health conditions such as fibromyalgia, postural orthostatic tachycardia syndrome (POTS), functional gastrointestinal disorders and cardiovascular dysautonomia (Castori et al., 2017). Its multi-systemic nature creates challenges for those living with JHS and professionals involved in their care (Clark and Simmonds, 2011). Diagnosis is delayed by a lack of understanding of JHS amongst healthcare professionals (HCPs) (Grahame and Hakim, 2008; Baeza-Velasco et al., 2011; Palmer et al., 2016). Consequently, those with JHS often describe feelings of loneliness, marginalisation and isolation (Baeza-Velasco et al., 2011), with associated mental health issues (Smith et al., 2014; Martín-Santos et al., 2010).

Reliable prevalence figures in the general population are notoriously elusive due to differences in diagnostic criteria and a lack of high quality epidemiological research. However, between 30% (Connelly et al., 2015) and 55% (Clark and Simmonds, 2011) of people attending musculoskeletal clinics have been identified with JHS. It should be noted that the diagnostic criteria and nosology have recently changed, with the terms 'hypermobile Ehlers-Danlos Syndrome (hEDS)' and 'Hypermobility Spectrum Disorder (HSD)' now advocated (Castori et al., 2017, Malfait et al., 2017).

The current research pre-dated the new diagnostic terms and therefore 'JHS' will be used throughout.

Our knowledge of the impact of JHS comes from relatively limited literature. For example, Terry et al. (2015) explored the lived experiences of 25 people with JHS using focus groups, identifying the most challenging aspects of living with JHS as pain, fatigue, proprioceptive difficulties and recurrent injury. Participants described a lack of HCPs' and others' awareness of JHS, which contributed to tortuous routes to diagnosis and accessing appropriate healthcare services. Palmer et al. (2016) separately reported data related to physiotherapy management from the same 25 people with JHS, comparing this with data from 16 HCPs. Across both groups, participants agreed on the need for better knowledge of the condition and more recognition of the complex issues it causes. De Baets et al. (2017) interviewed 10 mothers with JHS, and found that acquiring a diagnosis was the starting point to organising their lives and managing their symptoms more effectively. It was reported that hypermobility caused emotional, social and physical problems, affecting activities of daily living such as cooking, shopping and walking. Similarly, Schmidt et al. (2015) used semi-structured interviews to investigate women's experiences of chronic pain in JHS. The unpredictable nature of the condition led to fear-avoidance of certain activities and carried a high emotional cost. It is clear that more qualitative research is needed to provide rich, high quality, in-depth data to inform practice, service delivery and patient-centred care (Hammel, 2004).

Physiotherapy is the mainstay of treatment for JHS (Palmer et al., 2016). However, few physiotherapists have received JHS-specific training, and so they are unfamiliar

with its presentation and lack confidence in its management (Lyell et al., 2015, Palmer et al., 2015, Rombaut et al., 2015). Physiotherapists also reported limited use of validated diagnostic and assessment tools (Palmer et al., 2015). To devise and monitor an effective management plan, physiotherapists and other HCPs need to both understand the impact of JHS on their patients and assess that impact. Patient-reported outcome measures (PROMs) are often used to identify and monitor the impact of health conditions. Recently, a condition-specific PROM was developed for people with JHS – the Bristol Impact of Hypermobility (BloH) questionnaire (Palmer et al., 2017a). As part of its development, a longer draft questionnaire was administered to adult members of the Hypermobility Syndromes Association (HMSA), a patient organisation in the UK. The draft questionnaire was accompanied by an already well-validated general health questionnaire, the Short-Form 36 (SF-36), which was previously shown to be sensitive to change following an exercise intervention in people with JHS (Ferrell et al., 2004). The questionnaire pack also included an open text box labelled: *“Any further comments you would like to add”*. 615 valid responses were received by Palmer et al. (2017a) and a quantitative approach was then used to select the items for the final questionnaire. The final BloH questionnaire items were subsequently found to exhibit strong concurrent validity with the SF-36 physical component score ($r=-0.725$, $n=615$, Palmer et al., 2017a) and, in a further study, excellent test-retest reliability ($ICC=0.923$, $n=233$, Palmer et al., 2017b). However, the qualitative comments gathered during the initial questionnaire development were not formally analysed. Anecdotal evidence suggested that they contained extensive information about patients’ experiences and the use of PROMs to identify those experiences.

This study aimed to explore the qualitative comments from previously administered questionnaires to identify (a) the impacts of living with JHS and (b) issues related to assessing those impacts using PROMs.

METHODS

A favourable ethical opinion was obtained from the South West 5 NHS Research Ethics Committee (reference 10/H0107/46).

Data handling and transcription

The methods of identifying and recruiting JHS participants were fully reported by Palmer et al. (2017a), Stage 3. Participants were members of the HMSA; had a self-declared diagnosis of JHS; were ≥ 18 years old; had no other formally diagnosed conditions affecting physical function (such as inflammatory arthritis, osteoarthritis or neurological conditions); were able to give informed consent; and were able to understand and communicate in English. Questionnaires were distributed by regular mail. A total of 636 questionnaires were received, of which 21 were excluded (12 were aged < 18 years; nine omitted BloH questionnaire items so a total score could not be calculated). A potential sample size of $n=615$ was thus available for the present study.

The research team included a very experienced senior investigator (SP) and five novice researchers (KB, IDP, RJ, CP and MW) who were final year physiotherapy students. Questionnaires had been allocated sequential participant numbers by the previous study team (Palmer et al., 2017a). Questionnaires were anonymised by

detaching demographic information and storing it separately, with the only remaining identifier being participant numbers.

Questionnaires were allocated to the novice researchers for transcription. Qualitative responses were transcribed verbatim into a Microsoft Excel spreadsheet, alongside the relevant participant number. Any identifiable data was further anonymised during transcription, for example by using '[name]' to replace a person's name. Unclear words were verified by other team members. Transcription was overseen by the senior investigator.

Analysis approach

Transcripts were analysed using thematic analysis, where researchers identified concepts and patterns of meaning in the data (Braun and Clarke, 2013). This process included complete coding, sub-theme and theme formation, and continuous discussion and critique amongst the research group. Thematic analysis was chosen as it is non-prescriptive and flexible, providing guidance but allowing the process to evolve with the data. It is also quick to learn, which benefitted the novice researchers (Braun and Clarke, 2013).

Coding

Braun and Clarke (2013) defined coding as examining the data and selecting all material relevant to the research question. Codes included distinct words or phrases, which revealed different concepts, issues and ideas. The coding process is illustrated in Figure 1. Initial coding was conducted on each data item by the same researcher who had transcribed it, thereby enhancing familiarity with the data. A

second coder was then then assigned to each data set and undertook blind double-coding. Using multiple analysts in this way has been shown to increase inter-rater reliability (consistency of analysis), rigour, quality and breadth (Curry et al., 2009, Pope et al., 2000).

Each pair of researchers then met to debate their codes, combining these to form one coding list per data set. Where disagreement occurred, the entire research group discussed the issue and came to a consensus. Resultantly, five coding lists were compiled, blanketing all questionnaire responses. The research group then scrutinised and debated these to form one final coding list. This extensive engagement in critical discussion surrounding codes further enhanced the quality of data analysis (Curry et al., 2009).

Next, the total dataset was divided in half and two pairs of researchers applied the final coding list to the raw data. This critical use of the coding list enhanced data analysis (Dierckx de Casterlé et al., 2012), and ensured that every questionnaire response was accounted for. A fifth researcher acted as an impartial third party, making the final decision in the case of disagreements. Researcher roles were allocated randomly.

Sub-theme and theme formation

Once the final coding of the data was confirmed, the group began to form sub-themes and themes from the data. Braun and Clarke (2013) defined themes as patterns of meaning which capture important points in the data. Themes can be made up of several smaller sub-themes, found by piecing together similar codes.

The process for sub-theme and theme formation is displayed in Figure 2. Each researcher printed and cut out the individual codes, then independently grouped these into sub-themes. Again, the group met and debated their sub-theme choices, using group consensus to make final decisions.

The data was again split into two sets and the same pairs of researchers from the coding stage applied the final sub-theme list to the raw data. Following this confirmation of sub-themes, the researchers independently collated the sub-themes into themes. The process of discussion, agreement and applying the themes to the raw data was repeated, until application of the final themes was complete.

Members of the research group kept reflective logs of their experiences and opinions throughout the data analysis process, as this has been shown to improve critical thinking and encourage observation, building more complex understandings of studied topics (Rich, 2015). The senior investigator regularly met with the researchers to discuss the analysis process and emerging findings.

RESULTS

One further questionnaire was excluded as the written comments revealed that it had been completed by an adult on behalf of their child (<18 years), although the parent had entered their own demographic details. A total of 614 eligible questionnaires were therefore available for inclusion. Of these, 393 (64%) contained written comments (with a mean of 63 words per respondent) and were analysed.

Respondents were slightly older than non-respondents (mean 41 and 37 years

respectively) but were comparable on the basis of gender (95% and 94% women) and BloH score (mean 228 and 227/360).

The final coding list contained 109 codes, which were grouped into 16 sub-themes and three main themes. The main themes were: 1) 'Impacts of living with JHS'; 2) 'Management strategies for JHS' and 3) 'Measurement and research into JHS.' The themes and sub-themes are illustrated in Figure 3 and described below. The full lists of themes, sub-themes and codes are provided as online supporting information. Participants are identified by sex and age. For example, '[F47]' is a 47 year-old female and '[M21]' is a 21 year-old male. '[F-]' or '[M-]' is used where age was not reported.

Theme 1: Impacts of Living with JHS

1a. Employment and education

Several responses described changes to working life due to JHS, for example one participant *"...retired from teaching early but trained as a reflexologist so that I could have control over the amount of time I worked each day at home..."* [F71]. Multiple participants referred to being *"...unable to work through disability"* [F40] or *"...trying to work from home so I can control my activity"* [F37]. Some participants described difficulties in education due to *"...pain in knees and hips, made worse during revision and exams due to long periods of sitting"* [F32]. However others reflected on more positive outcomes of their JHS diagnosis, such as *"...extra time @ school + uni + breaks – very helpful"* [F27].

1b. Mental wellbeing

Many participants described the impacts of JHS on their wellbeing, referring to problems such as feelings of depression, inability to cope and angst. One participant noted that JHS caused “...*stress, anxiety and frustration*” [F68] whilst another described the condition as “*detrimental to living a full life*” [F28].

1c. Diagnosis

Participants described both positive and negative impacts of JHS diagnosis, which was commonly a lengthy process. Some described the ways that diagnosis had helped them to manage: “*The diagnosis for me was life changing*” [F32]; “*Since being diagnosed + prescribed medications & physio + pilates I feel more in control of pain management. I have much more time feeling better/ stronger than I have all my life*” [F45]. Many participants stressed the importance of early diagnosis: “*Medical diagnose [sic] at an earlier time would have a huge impact on management of hypermobility for the better*” [F48]; “*I do feel bitter that it took until I was 50 to be diagnosed with HMS [hypermobility syndrome]. I had over 10 years of pure hell and feel if I had been diagnosed, all would have been easier*” [F51].

1d. Co-morbidities

Many of those responding to the questionnaire mentioned living with various co-morbidities. These included POTS, fibromyalgia and spinal pathologies, and can reportedly make it “...*hard to tell what illness is causing a problem*” [F18]. Some participants noted having mental health problems that interfere with symptoms, for example “*I also have bi-polar disorder and feel my hypermobility is affected by my moods...*” [F39].

1e. Physical complications of JHS

Participants highlighted various physical issues related to JHS, such as pain, fatigue, joint dislocations and subluxations. *“The fatigue is worse than the pain. If I walk a lot or work all week, I have to rest all weekend [...] it’s not a fatigue you can sleep off”* [F45]. Some participants *“...dislocate limbs often...”* [F56], and *“...suffer from migraines and headaches...”* [F50]. One participant reported that *“it isn’t always that I can’t do things – I don’t have the energy”* [F59].

1f. Social impact of JHS

JHS can impact a person’s social life too, causing problems such as social isolation, financial problems and putting strain on relationships. Some of the causal factors seemed to be tiredness, *“In terms of social life- I’m so tired by the end of the day that I rarely go out socialy [sic]”* [F21], and fear-avoidance of activities *“I have stopped my hobbies and visiting friends because I am worried I will hurt myself all of the time”* [F34]. Social isolation sometimes comes from misunderstanding of the condition *“friends/ family tend to leave you out of activities [sic] if they think you will not be able to take part even though they do not ask, this has on several occasions left me feeling isolated”* [M24].

1g. Nature of JHS

Multiple participants spoke of the changing nature and unpredictability of JHS: *“Hypermobility syndrome is such a variable beast according to your general wellbeing, physical demands and work commitments meaning that one 7 day period can be very different to the next”* [F44]; *“No one day is ever the same. You never know what part of your body is going to be in pain!”* [F52]. Its apparent invisibility

frustrated many people: *“People don't know how hard it is, because I look healthy”* [F50].

1h. Lack of JHS services and HCP knowledge and understanding

As identified in the literature, many with JHS can relate to feeling misunderstood and mistreated, causing negative experiences of the healthcare system. Participants described *“...a complete lack of understanding of HMS [...] with a range of medical practitioners”* [F40] and *“...a wide gap amongsts [sic] health care professionals understanding of the impact HM [hypermobility] has on a person's day to day function”* [F48]. One reported that *“...local doctors and rheum [sic] don't understand the problems so have no advice/ solutions”* [F32] and another that *“...normal physio advice to 'stop where it hurts' is often too late!”* [F57].

Theme 2: Management strategies for JHS

2a. Physical activity

Many use exercise, active lifestyle choices or other activities to manage their JHS. One female participant wrote: *“...by walking 1 hour a day and swimming twice a week, I feel more mobile and my joints feel 'better' supported and strong”* [F47]. One used *“...yoga for core strength and for breathing exercises to control pain and to relax”* [M48]. Another reported that they *“...try to keep my weight under control to ease strain on my joints, go to gym with a P.T. [personal trainer] [...], go to pilates and physio...”* [F35]. Not all comments surrounding exercise were positive, for example *“my muscles are very painful all my life I had done isometric exercises to try and keep them strong, but now it does not work”* [F66], and *“I would like to be more*

physically active but feel unable to do that because of the pain or injury I might suffer” [F53].

2b. Equipment

Various modes of supportive equipment were mentioned for managing symptoms of JHS, for example wheelchairs, mobility aids and orthotics. One participant noted being “...*reliant on walking aids and pain medication*” [F40]. Another reported that “*Every joint is now supported with splints/ brace [sic] and I am now coping a lot better*” [F57].

2c. Medication

Medication for sleep, pain and mental health conditions were noted, with a mixture of positive and negative accounts of their use and effectiveness. One “...*can only function when taking pain killer medication. I am taking anti-depressants, or my mood would be mush [sic] more down*” [F62]. Another described pain medications as ineffective, for example “...*even Tramadol (v. [sic] hard to get from GP!) only takes edge off back/knee pain when it's severe and leaves me too spaced out to work/do anything!*” [F50]. Another participant said “*it has taken a year to get my medication at a level which manages my pain*” [F30].

2d. Positive health care interventions

Healthcare interventions discussed included pain management courses, physiotherapy, adjunct and alternative therapies. One woman's back pain was “...*transformed by going to podiatrist (privately) & paying £500 to get proper insoles to support my arches in my feet*” [F58]. Another respondent said: “*I have been on a*

number of pain management courses and have managed to stay in work due to the coping strategies taught" [F41]. Others found cognitive behavioural therapy (CBT) helpful *"I feel much more positive about managing things and using CBT methods to manage pain"* [F39].

2e. Personal management strategies for JHS

Several comments were made surrounding people's pacing techniques and lifestyle adaptations to manage their JHS. Participants discussed the importance of positive attitudes and strong support networks for coping, such as the HMSA which one person described as *"...such an important association helping to support people and giving understanding of HMS"* [F35]. People noted that management of JHS is *"...a constant balance of not over-doing or under-doing it!!"* [F37] and accepting that *"there are things I cannot do and ether [sic] find other ways of achieving the same time [sic] or getting help - no point being upset or frustrated"* [M48]. Some acknowledged that finding this balance is difficult: *"I am the sort of personality which would push myself to do everything [...] so as to not let people down to the detriment of my health rather than pace. I will overstretch myself for weeks on end and then crash!"* [F37].

Theme 3: Measurement and research into JHS

3a. Issues with completing the questionnaire

Participants experienced various problems completing the questionnaire, related to the variability of JHS, difficulties with handwriting, and mental health impacting their responses. *"The condition is extremely variable making questionnaires rather difficult to gage [sic] and [sic] accurate portrayal of individual symptoms"* [F25]. *"An*

electronic version of this form would have been helpful as it is painful to use a pen!"
[F39].

3b. Feedback on questionnaire questions

Some participants suggested improvements. They commented on the ambiguous nature of certain questions, the need for more answer options and the need for questions on different areas of the body or aspects of JHS. *"Some answers would benefit from having a 'not applicable' option [...] Questions relating to other conditions that are related to hypermobility/ EDS would be helpful"* [M39]. Some expressed their satisfaction with the questionnaire: *"Well designed and asks appropriate questions regarding not just the physical but also the emotional aspect of the condition which is often overlooked or simply ignored"* [M23].

3c. Importance of JHS research

Many people voiced their appreciation for the research taking place and expressed a willingness to participate in further studies. *"Thank you for researching an area so poorly understood"* [F39]; *"Thank you for running this study. Hopefully more research into hypermobility will raise awareness of the condition and help those affected to manage pain/fatigue better"* [F44].

DISCUSSION

The study successfully identified data relevant to the impacts of living with JHS and issues related to assessing those impacts using PROMs. Participants also

commented on strategies for managing their condition, providing valuable insights into potentially effective lifestyle adaptations and therapeutic strategies.

Reported impacts of JHS

Many participants mentioned physical complications and pain was common in everyday life, as previously noted in the literature (Clark and Simmonds, 2011, Palmer et al., 2016, De Baets et al., 2017). Pain may be due to subluxations, tendinopathies, incongruent articular surfaces, proprioceptive deficits, nervous system sensitisation, and psychological inputs to the brain-self neuromatrix (Hakim et al., 2010).

Pain may be managed in several ways, for example through targeted drug therapy (Bird, 2010) and holistic long-term physiotherapy (Palmer et al., 2016). Baeza-Velasco et al. (2011) reported that CBT and a multi-disciplinary approach offer the best management for chronic pain in JHS. This can be provided through pain management programmes, as noted by many of the present study's participants. Baeza-Velasco et al. (2011) described promising results of a JHS pain management course that focused on recovering lost function, improving self-efficacy, activity pacing and reducing the need for analgesia. Further research and development in this area could benefit JHS management.

Participants also reported issues with mental and social wellbeing linked to JHS. Several noted feelings of distress, depression, social isolation and a lack of understanding and support. Smith et al. (2014) found that those with JHS experienced significantly more fear, anxiety, depression and panic disorders than

those without. Although this link is not necessarily causal, it reaffirms the need for a holistic biopsychosocial approach to management. Kostova et al. (2014) reported that supportive social environments facilitate acceptance for those with chronic pain, which can in turn improve quality of life and reduce pain and depression. Martín-Santos et al. (2010) stated that professionals caring for those with JHS and mental health issues must thoroughly understand the implications of their problems and how to appropriately address them.

Respondents conveyed the heterogeneous, unpredictable and variable nature of JHS, describing its wide-ranging impact and the lack of control they often feel over its management. This has resonance with previously reported findings that everyone with JHS has different symptoms (Palmer et al., 2016). Participants voiced frustrations over the invisibility of the condition, which exacerbates a lack of understanding and support from others. The nature of JHS presents several challenges. Many described having to change their activities of daily living and develop personal coping mechanisms. De Baets et al. (2017) also identified difficulty managing energy levels to carry out usual activities, such as walking or gardening. Participants in that study discussed management options such as pacing, taking naps and prioritising certain activities over others (De Baets et al., 2017). In the current study, participants also commented on the importance of pacing activities, knowing their own limits, and joining supportive groups such as the HMSA. It is clear there is not a single solution to the many problems that JHS imposes, however there are options available when developing management plans.

Many participants noted difficulties during their journeys to diagnosis. Some reported that HCPs disregarded their symptoms or failed to use the correct diagnostic tools. It has been well-documented that diagnosis of JHS is a difficult and convoluted process (Baeza-Velasco et al., 2011; Terry et al., 2015) and that this can lead to misunderstanding and poor outcomes for patients (Grahame and Hakim, 2008). Addressing this, the HCPs studied by Palmer et al. (2016) highlighted the need for more sensitive diagnostic criteria. Therefore, it will be interesting to observe any impact of recent changes to the diagnostic criteria (Castori et al., 2017; Malfait et al. 2017).

A lack of understanding and support for those with JHS was evident. Many participants described encounters with HCPs and others in which they did not feel heard. One potential explanation offered by other authors (Rombaut et al., 2015; Terry et al., 2015; Palmer et al., 2016) and by this study's participants, is the lack of JHS-specific training for HCPs. Practitioner education may enhance outcomes for those with JHS. Palmer et al. (2017a) developed the BloH questionnaire to assess the specific impacts of JHS. This tool could help HCPs and service users to identify and agree problem lists, create targeted treatment plans and work towards shared management goals.

Difficulties with assessing the impact of JHS

The development of the BloH questionnaire (Palmer et al., 2017a) has offered some advances in the assessment of JHS, with physiotherapists and service users commenting positively on its appropriateness, acceptability and feasibility and recognising its benefits for supporting JHS management (Manns et al., 2018).

However, assessing the impact of JHS using PROMs remains complicated. For example, in the current study, participants commented on the need for medication and equipment-related questions. Respondents noted finding it difficult to answer certain questions, for example those about mobility or pain, without considering which piece of equipment or dose of analgesia they might use. Exploring the current literature, it is apparent that no studies have explored the impact of specialist equipment on the lives of people with JHS. Rombaut et al. (2011) did, however, investigate the effects of drug treatment on pain severity and functional impairments in JHS patients. They found that 92.4% of participants used medication for their symptoms. Interestingly, high analgesia consumption was associated with higher levels of dysfunction, although the cause-effect relationships are unclear. This highlights the need for further research into the use of medications to relieve the symptoms of JHS.

Respondents also commented on the seven-day timeframe used in the draft BloH and SF-36 questionnaires. Some participants noted that this was not necessarily representative of their experiences in general, and so may not have provided an accurate overview of their experiences. The final published BloH questionnaire kept the seven-day timeframe (Palmer et al., 2017a) and therefore this is important to consider in its interpretation.

Participants also highlighted the need for their co-morbidities to be considered, such as POTS, fibromyalgia and osteoarthritis. JHS rarely exists in isolation (Castori et al., 2017) and so the sample studied was likely to be representative. Hakim and Grahame (2004) found that those with JHS in conjunction with non-musculoskeletal

symptoms were likely to experience more anxiety, fatigue and issues with sleep. Castori et al. (2017) explained that such complications may result from autonomic dysfunction and the side-effects of medications. In any case, these conditions should be taken into account when assessing JHS, since best treatment outcomes may be achieved when a holistic approach is taken (Palmer et al., 2016).

Many of the questions in the draft BloH questionnaire that were highlighted as being problematic (questions 32-35, 36-62 and 87-90) were subsequently excluded from the final BloH questionnaire. In total, 22 of the 35 questions specifically identified were excluded because they were scored as relatively unimportant and/or were highly correlated with other questions (Palmer et al., 2017a). This suggests that the process of item reduction largely addressed the issues highlighted in the qualitative comments. It should be noted, however, that only 12 participants (3% of respondents) included comments specifically related to the BloH questionnaire. This may indicate general satisfaction, as suggested by previous qualitative evaluation (Manns et al., 2018).

Strengths and limitations

Morse (2015) described how different researchers' skills and ideas can impact on the coding of data, which in turn can affect the dependability of study findings. However, highly rigorous methods of analysis were employed in the current study, including blind double-coding, independent parallel formulation of sub-themes and themes, and stringent procedures for discussion and consensus. The researchers made every attempt to work with a neutral stance, using complete coding and thematic analysis to uncover every possible finding in the data, without looking for particular

codes, sub-themes or themes. The coding list, sub-themes and themes were applied to the raw data several times to ensure that every participant was accounted for, and that the findings reflected their comments. Generalisability is not usually an important or desired trait in qualitative research, due to the often small and focused populations studied (Leung, 2015). However a large sample provided comments in the present research (n=393) and they are likely to be representative of the wider sample (n=615) reported by Palmer et al. (2017a). 95% of respondents were female in both samples and the mean age and BloH questionnaire score was 41 years and 228/360 in the present sample (versus 40 years and 234/360 in the wider sample). Recruitment was via a patient organisation and therefore participants may differ from the wider population of people with JHS in terms of their knowledge and understanding of their condition and their motivations for taking part in the research.

Anderson (2010) stated that qualitative research is highly dependent on the skills of the researchers, and can be influenced by personal bias. Although the researchers who conducted analysis were novices, this reduced potential bias as they had little knowledge related to JHS. The large sample size of collected data resulted in a lengthy process of transcription and analysis, and made it difficult to gain an overall consensus or conclusion due to the high volume of different responses. Thematic analysis, however, allowed the researchers to effectively identify and compile the key themes. This qualitative study has provided a wealth of useful information to inform practice and future assessment of the impacts of this condition.

CONCLUSION

This project has illustrated the lived experiences of adults with JHS. Impacts were wide-ranging, including problems with joint dislocation, fatigue, social isolation and mental wellbeing. Medication and equipment to support living with JHS were two areas identified which have not previously been investigated in detail and could inform healthcare practice. There is a substantial need for education about JHS, both for HCPs and the general public. Such education could improve the quality of care and reduce stigmatisation. Specific feedback was given on the SF-36 and draft BloH questionnaires administered as part of this research, which could inform future adaptations or updates. There was much data related to the management of JHS, with each person managing their symptoms in vastly different ways. The challenges are many and varied, but with a holistic and multidisciplinary approach and well-equipped HCPs, the recognition and management of JHS could be improved.

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FIGURE LEGENDS

Figure 1. Flow chart depicting the process of transcription and coding.

Figure 2. Flow chart depicting the process of thematic analysis.

Figure 3. Final themes and sub-themes.

Figure 1. Flow chart depicting the process of transcription and coding.

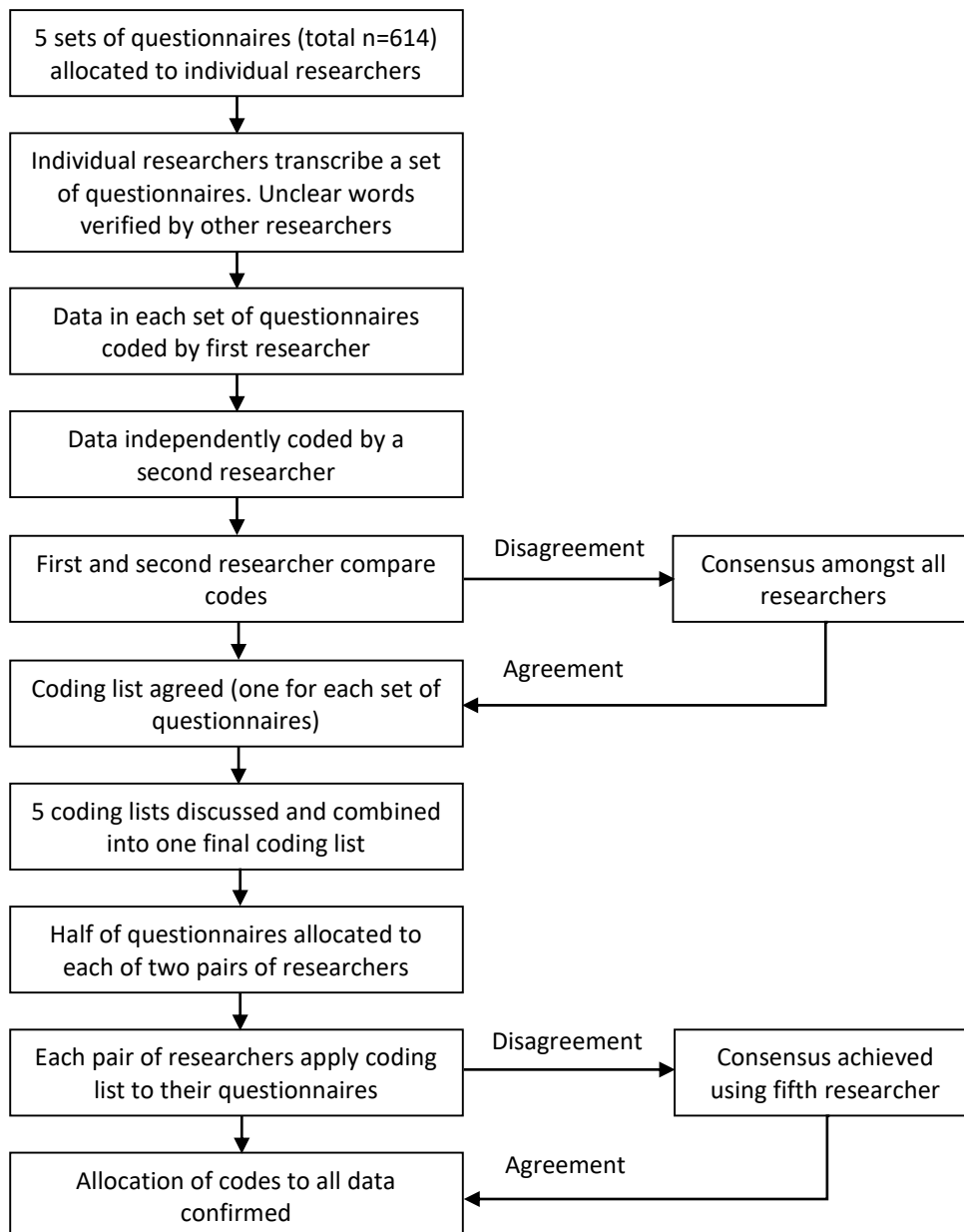


Figure 2. Flow chart depicting the process of thematic analysis.

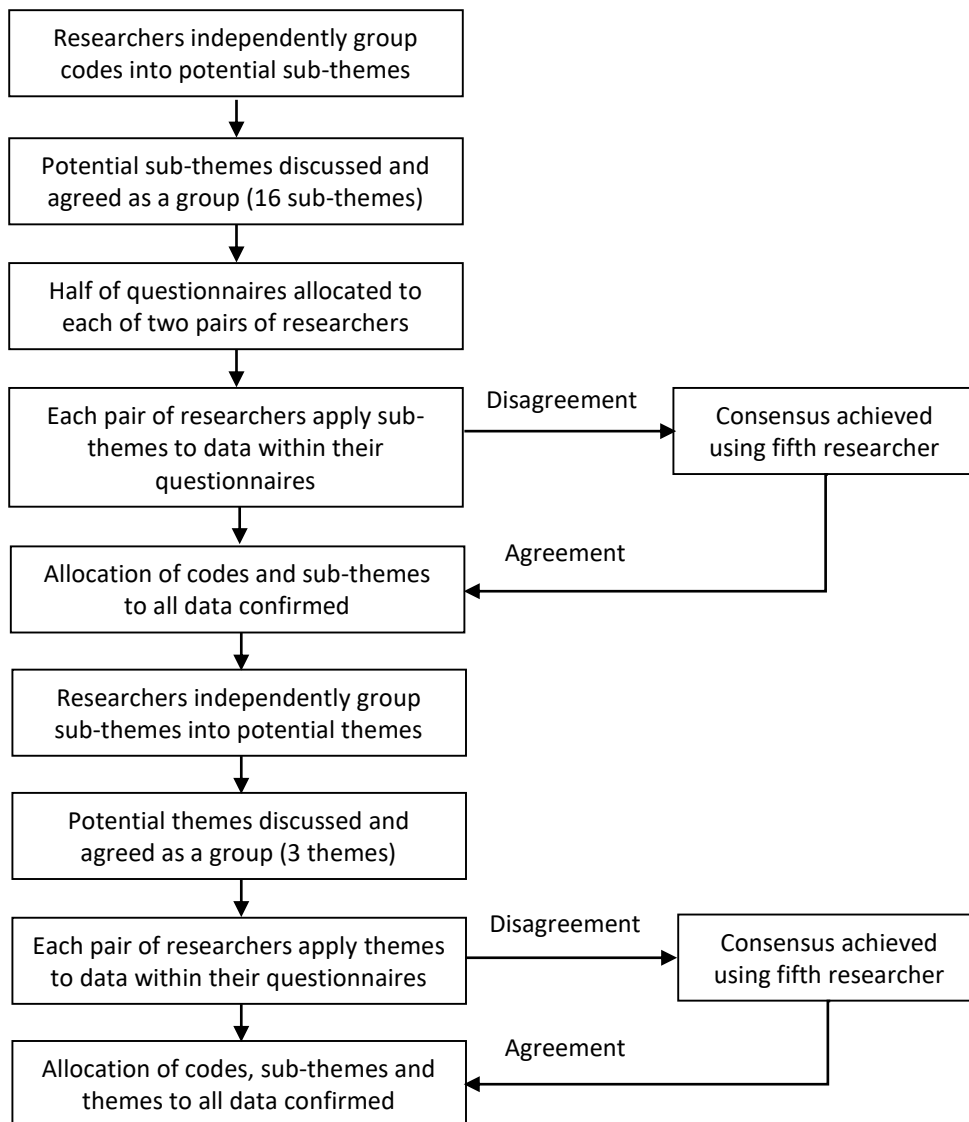
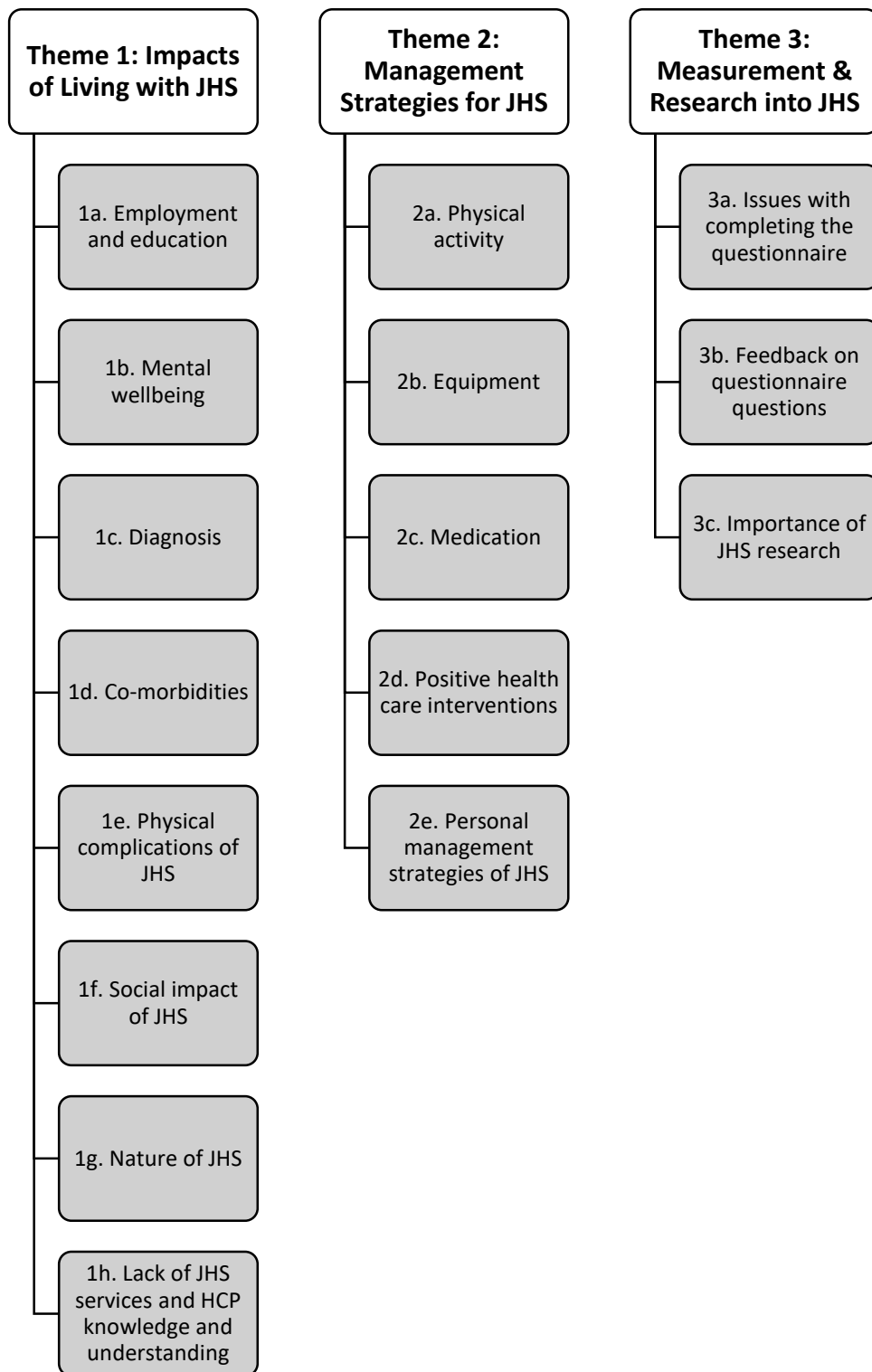


Figure 3. Final themes and sub-themes.



SUPPORTING INFORMATION

COMPLETE LIST OF THEMES, SUBTHEMES & CODES

Theme 1: Impacts of Living with JHS

- 1a. Employment and education
 - Difficulty managing employment
 - Lack of employer understanding and support
 - Unable to work due to JHS
 - School/college education impacted
- 1b. Mental wellbeing
 - Negative impacts of JHS on mental health
 - Frustration of having JHS
 - Inability to cope
- 1c. Diagnosis
 - Delay in diagnosis
 - Negative impacts of JHS diagnosis
 - Positive impacts of JHS diagnosis
- 1d. Co-morbidities
 - Low blood pressure
 - Arthritis
 - Fibromyalgia
 - Co-morbidities mentioned
 - Postural Orthostatic Tachycardia Syndrome
 - Dysarthria
 - Spinal surgery
 - Disc pathologies
- 1e. Physical complications of JHS
 - Long flight caused fatigue and discomfort
 - Dislocations and subluxations due to JHS
 - Lower back pain
 - Gastrointestinal symptoms
 - Symptom of pain
 - Temporo-mandibular joint problems
 - Neck pain
 - Symptom of fatigue
 - Pre-menstrual cycle impact on JHS
 - Skin fragility
 - Impact of pregnancy/childbirth on JHS
 - Clumsiness
 - Increased injury risk
 - Difficulties/pain with hand writing
 - Seasonal impact on symptoms
 - Sleep disturbances
 - Impacts of JHS on mobility
- 1f. Social impact of JHS:
 - Lack of general support
 - Lack of general knowledge and understanding of JHS
 - Social isolation

- Impact on intimacy
 - Lack of access to financial support
 - Stigmatisation and discrimination
 - Limitations to activities of daily living
- 1g. Nature of JHS
- Variability of JHS
 - Invisible nature of JHS
 - Multiple joints affected by JHS
 - Hereditary nature of JHS
 - Progression of JHS
 - Unpredictability of JHS
 - JHS is a disability not a health issue
 - Difficult to separate symptoms of JHS and co-morbidities
- 1h. Lack of JHS services and HCP knowledge and understanding
- Lack of HCP knowledge and understanding of JHS
 - Lack of service provisions for JHS
 - Negative physiotherapy interventions
 - More specialists needed in JHS management

Theme 2: Management Strategies for JHS

- 2a. Physical activity
- Walking to manage JHS
 - Positive impact of exercise
 - Pilates and yoga
 - Active lifestyle
 - JH aided gymnastics
 - Challenges of exercise
- 2b. Equipment
- Wheelchair use
 - Mobility aid use
 - Supportive footwear
 - Splints and orthotics
- 2c. Medication
- Medication for sleep
 - Medication for pain
 - Medication for mental health
 - Negative experiences of medication
 - Positive experiences of medication
- 2d. Positive health care interventions
- Positive impact of physiotherapy
 - Pain management
 - Interventions for mental wellbeing
 - Podiatry input
 - Acupuncture
 - Osteopath involvement
 - Alternative/adjunct therapies
- 2e. Personal management strategies of JHS
- Importance of maintaining independence
 - Overall good health
 - Importance of pacing

- Difficulties pacing
- Lifestyle adaptations required
- Impact of diet
- Coping strategies
- Importance of weight management
- The need to self-educate and self-manage
- Good support network
- Importance of positive attitude

Theme 3: Measurement & Research into JHS

3a. Issues with completing the questionnaire

- Variability of JHS affects responses
- Mental health affects responses
- Difficulties completing questionnaire due to hand pain from writing

3b. Feedback on questionnaire questions

- Include temporo-mandibular joint questions
- Include more answer options/ N/A option/ scale
- Include equipment questions
- Some questions too ambiguous
- Questions 32-35 too ambiguous
- Questions 36-62 too ambiguous
- Questions 87-90 too ambiguous
- Include medication question
- Include hobbies
- Include more upper limb questions
- 7-day timeframe of questions
- Include question about co-morbidities
- Questionnaire not a true reflection of JHS
- Questionnaire scores indicate ability to manage JHS
- Recognising own limitations following completion of questionnaire

3c. Importance of JHS research

- Appreciation of research into JHS
- Hopeful for further research into JHS
- Willing to participate in future research
- Recommendations for future research