Proprioception and muscle torque deficits in children with hypermobility syndrome

F. Fatoye¹, S. Palmer², F. Macmillan³, P. Rowe⁴ and M. van der Linden³

Objectives. Sensorimotor deficits such as impaired joint proprioception and muscle weakness have been found in association with hypermobility syndrome (HMS) in adults. HMS is more common in children than adults, yet such deficits have not been adequately investigated in paediatric populations. It is therefore uncertain as to what sensorimotor deficits are present in children with HMS. This study investigated knee joint proprioception and muscle torque in healthy children and those with HMS.

Methods. Thirty-seven healthy children (mean age ± S.D. = 11.5 ± 2.6 yrs) and 29 children with HMS (mean age ± S.D. = 11.9 ± 1.8 yrs) participated in this study. Knee joint kinaesthesia (JK) and joint position sense (JPS) were measured, with the absolute angular error (AAE) calculated as the absolute difference between the target and perceived angles. Knee extensor and flexor muscle torque was assessed and normalized to body mass. Mann–Whitney U-tests were performed to compare JK, JPS and muscle torque between the two groups.

Results. Children with HMS had significantly poorer JK and JPS compared with the controls (both P < 0.001). Knee extensor and flexor muscle torque was also significantly reduced (both P < 0.001) in children with HMS compared with their healthy counterparts.

Conclusions. The findings of this study demonstrated that knee joint proprioception was impaired in children with HMS. They also had weaker knee extensor and flexor muscles than healthy controls. Clinicians should be aware of these identified deficits in children with HMS, and a programme of proprioceptive training and muscle strengthening may be indicated.

Key words: Hypermobility syndrome, Joint kinaesthesia, joint position sense, muscle torque, proprioception, sensorimotor deficits.

Introduction

Generalized joint laxity in children is known to be associated with musculoskeletal pain [1] and its prevalence has been reported to be as high as 64.6% [2]. It reduces with increasing age [3] and girls are more affected than boys [3–5]. Generalized joint laxity may, however, present no ill effects to some children [6] and it may actually be an advantage in some sports, such as ballet dancing [7].

Hypermobility syndrome (HMS) is diagnosed when generalized joint laxity becomes symptomatic with musculoskeletal complaints in the absence of signs of any rheumatic, neurological, skeletal or metabolic disorders [8]. Joint pain is the major presenting complaint in children with HMS [9] and was found in 92 (74%) out of 125 HMS cases investigated by Adib et al. [10]. The symptoms of HMS have been demonstrated to be most common in the knee joint [9–11].

Proprioception deficits at the proximal interphalangeal [12] and knee [13, 14] joints have been found in adults with HMS. Muscle weakness has also been observed in children [10] and adults [15] with HMS. However, other studies have revealed no significant difference in total muscle torque measurements between healthy children and those with HMS [6, 8, 16]. Recently, musculoskeletal reflex dysfunction was also identified in adults with HMS [17].

To date, no published studies have investigated knee joint proprioception in children with HMS, with previous studies focusing instead on adults [13, 14]. Although muscle performance has been studied in children with HMS [6, 8, 16], the muscles acting on the knee have not been examined. In addition, previous sample sizes have been small (range = 13–19 participants [6, 8, 15]), making it difficult to generalize the findings. Therefore, the extent of sensorimotor deficits affecting the knee joint in children with HMS remains unclear.

Researchers have suggested that HMS is poorly recognized and that a long delay in accurate diagnosis results in poor management of its symptoms [14]. This may lead to disruption of childhood activities, such as school and play [10]. In order that children with this condition receive appropriate treatment, there is a need for clinicians to understand the factors associated with HMS. Hence, the purpose of this study was to compare knee joint proprioception and muscle torque in children with HMS to their healthy counterparts.

Methods

This report is part of a larger trial investigating a wider range of impairments, activity limitations and participation restrictions in children diagnosed with HMS. The proprioception and muscle torque impairment data are reported separately here to allow full exploration of the sensorimotor consequences of HMS in relation to the relevant literature.

Participants

Sixty-six children (37 healthy and 29 diagnosed with HMS) aged 8–15 yrs participated in this study. The mean age ± S.D. of the healthy controls (20 boys and 17 girls) and HMS group (8 boys and 21 girls) was 11.5 ± 2.6 and 11.9 ± 1.8 yrs, respectively. Exclusion criteria included a diagnosis of Ehlers–Danlos syndrome (EDS), musculoskeletal disorders, a history of trauma to either knee joint, or vestibular system disorders. It is believed that EDS type III and HMS are the same condition [7]. Three children with EDS were excluded, however, as the precise EDS diagnosis was unavailable from their history. The study was approved by the City of Edinburgh Council Education Department, Queen Margaret University (QMU) Ethics Committee and the NHS Lothian Local Research Ethics Committee. Healthy children were recruited from local schools in Edinburgh through their head teachers. Children with HMS were identified using the medical record systems at the Rheumatology Department at the Royal Hospital for Sick Children in Edinburgh and at the Podiatric Clinic, Springwell House in Edinburgh (over the period 2000–05).
All children identified in this manner were approached by letter. Following receipt of information regarding the study, both parents and children gave written consent before participation in the study. A sample size calculation (at 90% power and \( \alpha = 0.05 \)) was performed based on a pilot study conducted on healthy children. The calculation was based on two independent group comparisons (assuming normality) with a minimum clinically relevant difference of 3° and an s.d. of 2.7° in knee joint position sense (JPS). A minimum sample size of 27 children in each group (healthy children and those with HMS) was estimated on this basis.

**Testing procedure**

The first author (F.F.) undertook all experimental procedures. Following a brief explanation of the procedure, a diagnosis of HMS was confirmed in the HMS group on the basis of a Beighton score [18] of \( \geq 6 \) in the presence of multiple joint pain (presently or historically). The Brighton criteria now seem to be more widely recognized for diagnosing HMS but were not employed in the present investigation as they have yet to be validated in children. The most painful knee as reported by the children with HMS was tested. The test knee (i.e. left or right) of the healthy participants was determined using computer randomization. Data collection included knee joint kinaesthesia (JK), JPS, and maximum isometric flexor and extensor muscle torque. Since it is known that exercise training may enhance joint proprioception [19, 20] and muscle torque [8], the level of physical activity was documented as the amount of time (hours) spent on sports and/or physical education per week [8]. To facilitate the practicalities of testing, the order of specific tests was the same for all participants: JK, JPS, extensor muscle torque and finally flexor muscle torque. Testing took place in the Human Motion Analysis Laboratory at QMU Edinburgh.

**Joint proprioception assessment**

Knee joint proprioception was determined with a purpose-built motorized proprioception measuring device (parts supplied by Automated Motion Systems, Perth, Western Australia) using similar methods described by Barrack et al. [21], Corrigan et al. [22] and Grob et al. [23]. The device, previously validated [24], was constructed from a slow-speed motor mounted on a drive shaft, a motor control with a participant’s on/off switch, an inextensible belt and a pulley attached to the drive shaft. The inextensible belt was connected to a limb support attached to a frame that was made of a piece of aluminium rod and a protractor from which the angular displacement could be read.

**Joint kinaesthesia**

JK was assessed using threshold to detection of passive movement with participants in high sitting on an adjustable plinth with their back supported and reclined to 60° to encourage relaxation. Participants sat with their legs hanging freely over the edge of the plinth 4–6 cm proximal to the popliteal fossa, such that the knee joint was not in contact with the plinth [20]. Visual and auditory sensory inputs were eliminated by means of blindfold and earmuffs, respectively [25]. The test limb was positioned such that the rotation axis of the driving shaft of the device was in line with that of the participant’s test knee joint (lateral femoral condyle) and the lower leg was placed in a padded limb support (Fig. 1) [24]. Since joint receptors are involved in joint proprioception only when the muscle stretch receptors are not available [26], it was believed that proprioceptive input from muscle stretch receptors was minimized by placing the test limb in a limb support as this would help to achieve relaxation of the muscles acting on the knee joint. The starting position of the test knee was maintained at 60° of flexion [24].

The participant’s limb was then moved into extension at a constant angular velocity of 0.38°/s after a random delay of between 5 and 15 s [27]. Three practice trials were administered to familiarize participants with the test protocol. During the practice trials, the examiner pressed the response button immediately the test limb started moving. Participants were asked to relax and to mentally note a clear sensation of movement or change in position of their lower limb.

Thereafter, each participant was given the button and was instructed to press it once he/she detected position change in the test limb. To ensure that legitimate data were collected from each participant, three test trials were carried out where the second trial was used for data analysis [24].

**Knee JPS**

JPS testing can be performed under either weight-bearing or non-weight-bearing conditions. Although the former condition provides proprioceptive information under functional condition [28] it involves both motor and sensory receptors [29] and is more complex to test. Impaired muscle function [16] and pain [9] may be associated with HMS, and may affect the ability of children to perform a weight-bearing test. Passive JPS tests were therefore used for this study. This ensured that proprioceptive input emanated from the test joint and not from the muscles acting on the knee joint. The experimental set-up and participants’ position was identical to that described above for JK testing, except that the starting position of the test knee was 90° flexion [24].

The motorized device passively moved the participant’s test limb from the starting position (90° of knee flexion) to two pre-determined test angles (25° and 10° knee flexion), at a constant angular velocity of 2.2°/s [24]. Three practice trials at each test angle were administered, where the investigator stopped the device. This was followed by three test trials at the test angles when the participants pressed the response button themselves to stop the device. To enable the participant to identify and remember the target positions, the test limb was held in these angles for \( \sim 10 \) s [30] during the practice trials. Participants were asked to concentrate on the position. The leg was then returned to the starting position using the motor, where it was left for \( \sim 15 \) s [30] before the next practice or test trial.

Participants were instructed to relax during the test trials and were told to press the response button when they thought that the limb had reached the target position previously demonstrated. The absolute angular error (AAE), the absolute difference between target and perceived angle, at each test angle was calculated.
Muscle torque

Maximum isometric knee extensor and flexor muscle torque was quantified using a digital myometer (MIE, Medical Research Ltd, Leeds, UK). The experimental procedure has been described in detail by Fatoye et al. [24]. Participants sat on a plinth with their back supported and reclined to 60° and the test knee flexed to 90° (Fig. 2) [24]. Extensor muscle torque was measured first by instructing the participants to straighten their knees from the starting position [24]. Flexor muscle torque was then assessed, with participants being instructed to bend the test knee as much as possible.

Participants were given consistent verbal instruction and encouragement. One maximum contraction produced by both the extensors and flexors was assessed [displayed as force in Newtons (N) on the myometer]. Muscle torque was calculated as possible.

The characteristics of the participants are presented in Table 1. The median and interquartile range (IQR) of the Beighton scores for children with HMS was 7.0 (2.0). The median (IQR) of all measurements in both groups are illustrated in Table 2. The results of all the outcome measures based on gender are presented in Tables 3 and 4. Figure 3A–C illustrates the median and IQR for JK and AAE for the JPS measurements at 25 and 10°, respectively in the two cohorts. Figure 4A and B displays the median and IQR of knee extensor and flexor muscle torque in the two groups.

Results

The HMS group had higher median AAE values of JK and JPS tests than healthy children and these differences were statistically significant (P-values all <0.001). The median values of knee extensor and flexor muscle torque were higher in healthy children than in the HMS cohort and these differences were also statistically significant (both P<0.001). JK error and AAE for JPS tests were statistically significantly higher (P-range = 0.001–0.028) in girls with HMS than healthy girls. Similarly, JK error and AAE for JPS tests were statistically significantly higher (both P=0.001) in boys with HMS than healthy boys. In addition, knee extensor and flexor was statistically higher (P-range = 0.006–0.025) in healthy girls than in girls with HMS. However, muscle torque in boys with HMS was not statistically different (P-range = 0.309–0.360) from healthy boys.

Discussion

The lack of adequate knowledge of the symptoms associated with HMS in children makes the condition difficult to recognize.
and manage effectively [14]. To date, no studies were found investigating proprioception deficits in children with HMS. As a result, the findings of the present study on joint proprioception were examined in relation to the existing literature in adults. In the current study, knee joint proprioception (JK and JPS) was observed to be significantly poorer in children with HMS than their healthy counterparts. This observation is consistent with the findings of Mallik et al. [12] and Hall et al. [13] in adults with HMS.

The reasons for the identified proprioception deficits in the present study are not known. Furthermore, it is uncertain whether proprioceptive deficit in children with HMS is from birth or it develops during childhood. Children with this condition have been found with clumsiness and poor coordination [10] and adults with HMS have also been described as being clumsy in childhood [14].

It is believed that clumsiness and poor coordination may be related to poor motor development and impaired proprioception [10]. Infants with generalized joint laxity were found with delayed motor development [31, 32]. Generalized joint laxity is a prime feature of HMS [8] and poor muscle development has also been identified in children with HMS [33]. Since proprioceptive ability depends largely on sensory contribution from muscular receptors [34], it is possible that poor muscle development could contribute to proprioceptive deficit and poor motor development in children with HMS. Although motor development was not examined in the present study, this could serve as an area for future investigation.

Proprioception deficit has previously been attributed to diminished muscle spindle activation [34]. Muscle spindle sensitivity is controlled by the γ-motoneurone system and diminished activation of this system causes decreased muscle spindle sensitivity, resulting in proprioceptive impairment [34]. In the current study, passive JPS tests were administered, as this was thought to maximally stimulate slow-adapting joint mechanoreceptors while minimally stimulating muscle receptors [35]. In addition, the test limb of our participants was placed in a padded limb support during proprioception testing, as it was believed that this would enhance relaxation of the muscles acting on the test joint. It can, therefore, be assumed that proprioceptive contribution from muscle spindles was minimized. The knee joint proprioception deficit observed in children with HMS in the present study is therefore unlikely to be due to diminished muscle spindle sensitivity.

Knee joint proprioception deficit in children with HMS may also be due to diminished proprioceptive discharge from the joint receptors [12], although the mechanisms responsible for this are not well understood. Abnormal joint biomechanics due to joint laxity may lead to repetitive stresses on joints causing damage to the receptors [12]. Therefore, the diminished proprioceptive discharge from joint receptors may be related to damage to these receptors or defects in the articular tissues [12]. Moreover, knee joint proprioception deficit in children with HMS could be due to diminished joint proprioceptor activation resulting from capsular
or ligamentous stretching [12]. These factors may account for our finding but a causal relationship has not been established.

Proprioception can also be influenced by the menstrual cycle [36, 37]. For example, knee JPS deficits were observed in the menstrual phase [36] and impaired knee JK was detected during the pre-menstrual phase [37] in healthy women. In the present study, information regarding menstruation and age of menarche was not collated and therefore it is not possible to determine when measurements were taken in relation to the menstrual cycle. Group comparisons of JK and JPS data based on gender revealed that girls with HMS had significantly poorer ($P < 0.05$) knee joint proprioception compared with their healthy counterparts, just as observed for boys. Any effect of the menstrual cycle on knee proprioceptive acuity was therefore likely to be broadly comparable between healthy girls and those with HMS.

Extensor and flexor muscle torque was found to be significantly higher in healthy children than those with HMS, suggesting that children with HMS had muscle weakness. Presently, there is no available information regarding knee muscle torque in children with HMS as previous studies have focused on other joints [6, 8, 16]. These previous investigations failed to observe muscle strength deficits, perhaps because the joints they examined were asymptomatic. In addition, methodological differences between the present study and those of Engelbert et al. [6, 8, 16] limit detailed comparisons of the findings.

The reasons for the reduced muscle torque in children with HMS are not known. Laforte et al. [38] demonstrated that knee flexor and extensor muscle torque was enhanced by training. Adib et al. [10] found that the majority of children with HMS did not engage in routine exercise such as physical education at school. Therefore, the level of physical activity in children with HMS could have been responsible for the current findings on muscle torque. However, this explanation would seem unlikely as independent $t$-test analysis revealed no significant difference ($P = 0.740$) in the level of physical activity between healthy children and those with HMS.

The lower values of muscle torque recorded by the HMS group may instead be due to incomplete voluntary activation of muscles acting on the painful knee joint [39, 40]. Although not formally recorded as part of the current research, the majority of the children with HMS complained of knee pain during muscle torque measurements. It is possible, therefore, that the presence of pain during muscle torque testing may have impaired maximum muscle contraction, although this requires further investigation.

The implications of the findings of the present study are that impaired knee joint proprioception could lead to limitation of functional performance involving the neuromuscular system [41], such as walking [32]. Knee muscle strength has also been shown to be a determinant of lower limb functional activities [42] and impaired muscle function may limit activities of daily living (ADL) [43]. Therefore, proprioception and muscle torque deficits found in children with HMS in the present study may be associated with functional limitations in children with this condition. The relationships between these deficits and function require verification in future research in children with HMS.

Improved proprioception and increased muscle strength have been reported in adult patients with HMS following an 8-week closed kinetic exercise programme [14]. Supportive splints have been shown to improve knee joint proprioception in healthy adults [44] and those with knee arthritis [45]. Enhanced knee joint proprioception has also been observed with the use of elastic bandages in adults with knee arthritis [29] and healthy adults [46]. Furthermore, strength training has been found to increase hamstring and quadriceps muscle strength in healthy children [47]. Therefore, it seems likely that proprioceptive acuity and muscle torque can be improved in children with HMS with appropriate intervention methods.

Given the present findings, it may be worth examining proprioceptive acuity and muscle performance during clinical assessment of children with HMS. Improved knee joint proprioception and muscle torque may help to relieve the symptoms associated with HMS in children. Consequently, enhanced knee joint proprioception and muscle torque may also improve ADL performance in children with HMS. Therefore, clinicians may be aware of these identified sensorimotor deficits in children with HMS and develop appropriate rehabilitation programmes to address these deficits.

It is acknowledged that this study has some limitations. Only the knee joint was tested, thereby limiting the generalizability of the findings to other joints. Only HMS children aged 8–15 yrs were examined and therefore our results cannot be extended to younger or older individuals with this condition. The majority of children with HMS recruited to this study were girls, whilst there were more boys in the control group. The likely explanation for this is that girls are more affected with HMS than boys [9]. It is hoped that these limitations could be addressed in future studies.

This study is the first to investigate joint proprioception and muscle torque deficits in children with HMS. We demonstrated that children with HMS had sensorimotor deficits compared with the controls. This suggests that children with HMS may benefit from appropriate treatment methods of improving such deficits. Interventions to enhance knee joint proprioception and muscle strength should therefore be developed and tested in children with HMS.

### Rheumatology key messages

- Sensorimotor deficits were found in children with HMS.
- Proprioception enhancement and muscle strength training programmes may be of value to children with HMS.

### Acknowledgements

We thank the children and their parents who participated in this study. We are grateful to Sally Wilkinson (extended scope physiotherapist, Royal Hospital for Sick Children, Edinburgh, UK) for her contribution to the study concept and design and assistance in recruiting children diagnosed with HMS. Thanks are also extended to Robert Rush, Queen Margaret University, Edinburgh, UK for his statistical advice. Steve Kelly, Manchester Metropolitan University, UK is gratefully appreciated for his assistance with the schematic diagrams used in this article.

### Disclosure statement

The authors have declared no conflicts of interest.

### References
