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ORIGINAL ARTICLE

Prevalence and frequency of self-perceived systemic features in people with joint hypermobility syndrome/Ehlers-Danlos syndrome hypermobility type

Cliffton Chan[1] · Anne Krahe[1] · Yim Tang Lee[2] · Leslie L. Nicholson[1]

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Abstract

Some commonly reported systemic features of joint hypermobility syndrome (JHS)/Ehlers-Danlos syndrome hypermobility type (EDS-HT) are absent from nosologies due to insufficient validity. The primary aim was to examine the hypothesised high prevalence and frequency of orthostatic intolerance, easy bruising, and urinary incontinence in adults with JHS/EDS-HT and secondarily to determine the association between extent of generalised joint hypermobility (GJH) and these systemic features. A cross-sectional cohort study was conducted via online recruitment of medically diagnosed JHS/EDS-HT patients. A survey collected demographic data and clinical history. A subgroup of participants underwent physical testing of GJH using the Beighton score and Lower Limb Assessment Score (LLAS). Descriptive analysis was performed on demographic data and self-reported non-musculoskeletal systemic features. Correlation of GJH scores and systemic features were performed using Spearman's rank correlation. The survey was completed by 116 individuals (95% female; 16–68 years) with 57 (93% female) also participating in the physical assessment. The most prevalent systemic feature was orthostatic intolerance (98%), followed by easy bruising and urinary incontinence (97% and 84% respectively). Of those reporting symptoms of orthostatic intolerance, easy bruising, and urinary incontinence, 58%, 40%, and 18% described them as very highly frequent respectively (frequency > 75%). No significant correlations were found between the extent of systemic features and GJH scores as measured by either the Beighton score or the LLAS. The high prevalence and frequency of the systemic features found in this study, which are omitted in diagnostic classification criteria, suggest that further research on their diagnostic accuracy is warranted.

Keywords Bruising · Diagnostic · Gastrointestinal · Hypermobility · Orthostatic intolerance · Urinary incontinence

Introduction

Joint hypermobility syndrome (JHS) and Ehlers-Danlos syndrome hypermobility type (EDS-HT) are heritable connective tissue disorders, mainly characterised by generalised joint hypermobility, related musculoskeletal conditions, and fragile skin [1–3]. For both conditions, the diagnosis is made on clinical presentation in combination with medical and family history, as there are currently no confirmatory molecular or genetic tests [4, 5]. Despite having distinct diagnostic criteria (Brighton Criteria and Villefranche Criteria) [4, 5], JHS and EDS-HT have been widely recognised as the same clinical entity, due to the significant overlap of phenotypic features present in both conditions [6]. In this article, the term JHS/EDS-HT or syndromic hypermobility will be used. While the true prevalence of adult JHS/EDS-HT is unknown, it has been estimated to range from 1% [7] to 2% [8] with females more highly represented than males [9].

Since collagen synthesis is affected in people with JHS/EDS-HT, symptoms extend beyond the musculoskeletal system [7]. Health professionals, who are potentially unaware of the possible complex presentations of the condition, often fail to recognise the condition when patients present with chronic widespread pain and fatigue and seemingly unrelated symptoms such as dizziness and irritable bowel syndrome [10].

✉ Cliffton Chan
cliffton.chan@sydney.edu.au

[1] Discipline of Biomedical Science, School of Medical Science, Faculty of Medicine and Health, The University of Sydney, Sydney, Australia

[2] Discipline of Physiotherapy, Faculty of Health Sciences, The University of Sydney, Sydney, Australia

Individuals with syndromic hypermobility report that their condition is poorly understood and managed by health professionals [11, 12]. Frequently, they are "passed from pillar to post" and a correct diagnosis is often delayed and coincidental [11]. Management is often siloed with a patient consulting a gastroenterologist for their gastric issues, a cardiologist for their dizziness/syncopal episodes, and a physiotherapist for their migraines and fatigue while health professionals do not connect the symptoms. Indeed, in a survey of 251 JHS/EDS-HT patients in the UK, 52% had been on their diagnostic quest for over 10 years before receiving a correct diagnosis [13].

Receiving a timely and accurate diagnosis is crucial to facilitate effective management, thereby preventing and/or reducing the progression of chronic pain and fatigue, autonomic dysfunction, and psychosocial issues [14, 15]. In the absence of correct diagnosis, individuals with syndromic hypermobility report feeling stigmatised and isolated from society, leading to further decreased participation in activities and mental health issues [11]. Management of these patients, in particular physiotherapy intervention, has been reported to be less effective if diagnosis is delayed [11, 15]. Conversely, early and accurate diagnosis is shown to reduce health-related costs and medical resource uses (for example, medical imaging) in a similar multi-system disorder, fibromyalgia [16]. Timely and accurate diagnosis is likely to not only have a significant positive impact on the mental health of individuals with syndromic hypermobility, but would allow them to refocus their attention on treatment and sustainable management of their condition.

In the absence of genetic markers, the new diagnostic classification, proposed in 2017, aimed to better differentiate the hypermobile subgroup of Ehlers-Danlos syndrome from the other subtypes of the condition and other hypermobility disorders [17]. While more comprehensive than the previous diagnostic criteria for JHS (Brighton) and EDS-HT (Villefranche), the new criteria for hypermobile Ehlers-Danlos syndrome (hEDS) and hypermobile spectrum disorder (HSD) do not include many non-musculoskeletal features encountered clinically, reported anecdotally, and commonly described in the JHS/EDS-HT literature. The International Consortium acknowledges that this is due to their lack of established sensitivity and specificity [4, 5, 17]. Such features include, but are not limited to, urinary incontinence, orthostatic intolerance, and easy bruising [18–20]. These systemic features have the potential to significantly impact quality of life, physical functional capacity, recovery, and overall well-being [20, 21]. The 2017 Classification of Ehlers-Danlos syndrome recognises the presence and impact of these, suggesting that more research is needed to validate the association of these systemic symptoms with the condition [17].

A number of studies have examined and identified the association between the aforementioned systemic features with people diagnosed with JHS/EDS-HT [20–23]. However, few studies have investigated the prevalence or severity of these systemic features or their association with the extent of generalised joint hypermobility.

While disordered connective tissue is the likely cause of the most commonly recognised symptoms of the condition, namely joint hypermobility and consequent instability, what remains unclear is the relationship between generalised joint hypermobility (GJH) and other systemic features. That is, are the most hypermobile individuals those who experience the most frequent systemic symptoms? Since a deficiency in type I and III collagen affects the integrity of skin, ligament, muscular, and vascular tissue, our hypothesis is that individuals with more joint hypermobility would display greater frequency of systemic symptoms [23].

The primary objective of this study was to determine the prevalence and the frequency of self-perceived urinary incontinence, orthostatic intolerance, and easy bruising in an Australian adult cohort with medically diagnosed JHS/EDS-HT. The secondary objective was to determine if an association exists between the frequency of the self-reported systemic features and the extent of generalised joint hypermobility.

Materials and method

This study utilised a cross-sectional, observational study design.

Participants

Participants were recruited via advertising on two Australian JHS/EDS-HT patient support group websites (ConnecTed and EDS Australia) and by passive snowballing. Respondents over the age of 16 who had a diagnosis of JHS or EDS-HT from their treating physician were included in this study. Those respondents who had pre-existing diagnosis of another heritable connective tissue disorder (e.g. Marfan syndrome, Osteogenesis Imperfecta, Stickler's disease, or other subtypes of EDS) or a current orthopaedic condition that might restrict normal range of motion in their spinal and peripheral joints were excluded from the study.

Procedure

Participants who met the inclusion criteria were asked to complete a survey and invited to further partake in a single physical examination. All involvement was voluntary and participants provided informed consent prior to data collection. Ethical approval was granted by the University of Sydney's Human Research Ethics Committee (No. 2012/558).

The questionnaire collected information on age, gender, ethnicity, relevant clinical history (e.g. number of fractures and history of hernia), family history, and information relating

to the symptoms associated with JHS/EDS-HT. The 2107 Classification of the Ehlers-Danlos syndromes was not published at the time of data collection; however, it is anticipated that all participants would meet the criteria for either hEDS or HSD. Participants were asked whether they bruised easily (bruising following minimal trauma) and experienced urinary incontinence (loss of urine on coughing, sneezing, or exertion such as lifting/jumping) or orthostatic intolerance (dizziness when getting out of bed in the morning or when exercising in the heat or during/after a hot shower) via dichotomous question format requiring a "yes" or "no" response. An open space was provided for the option to provide further information about these problems. The frequencies of easy bruising, urinary incontinence, and orthostatic intolerance were surveyed using a 100 mm visual analogue scale. These three systemic features were investigated in this manner because they are relatively unidimensional, whereas gastrointestinal and cardiovascular features of the disease are multi-factorial and complex to quantify using this method of measurement and are beyond the scope of this study. One end was marked as "never" and the other was marked as "often", and participants were asked to rate how often they experienced each symptom by putting a mark on the relevant place on the lines.

Arbitrarily, a score of 0 was considered normal, a score between 1 and 25 mm was considered low frequency, a score between 26 and 50 mm was considered moderate frequency, a score between 51 and 75 mm was considered high, and 76–100 mm was considered very high frequency.

During the physical testing, participants were assessed using two generalised joint hypermobility tools: (i) the Beighton score (Appendix 1) [24], providing a total score out of 9, and (ii) the Lower Limb Assessment Score (LLAS) (Appendix 2) [25] providing a total score out of 12. Only measurements from one randomly allocated limb were recorded for the LLAS as a strong correlation between measurements from the left and right limb were previously reported [26]. All assessments were completed in the same order without prior warm-up and were conducted by musculoskeletal physiotherapists each having at least 10 years of clinical experience (CC, LN, and AK).

Specific Beighton score cutoffs, by Singh and colleagues (2017), were applied for the presence of generalised joint hypermobility for men and women of various age brackets as recommended by an Australian normative population study [27]. For the male participants, a cutoff of ≥ 4/9 was applied for ages ranging from 16 to 39 years and ≥ 2/9 for ages 40–59. A cutoff of ≥ 5/9 was used for females 16–39 years, ≥ 4/9 from 40 to 59 years, and ≥ 3/9 for ages 60–69. The Beighton score demonstrates good interrater reliability (ICC = 0.73) and good content validity [28, 29]. A score of ≥ 7/12 on the LLAS had been validated to identify generalised joint hypermobility in the adult population with a specificity of 86% and sensitivity of 68% and an excellent interrater reliability (ICC = 0.85) [26]. Therefore, a score of ≥ 7/12 on the LLAS was used as the cutoff point in this study.

Statistical analysis

Questionnaire and physical examination data were analysed using SPSS version 21 (IBM, NY, USA). The data were inspected for outliers and tested for normality using calculation of skewness. Descriptive analyses were performed on demographic data, and the prevalence and frequency of self-perceived non-musculoskeletal features. McNemar's test was used to determine the differences between the number of participants who met the Beighton score cutoffs and those who met the LLAS cutoff for identification of GJH. Correlations between the extents of hypermobility of all participants (as measured by the Beighton score and LLAS) and frequency of symptoms of systemic features (normal, low, moderate, high, or very high) were analysed using Spearman's rank correlation coefficient (ρ), with the level of significance set at a probability (p) value of less than 0.05. Spearman's rank correlation coefficient was used as non-parametric comparisons between ordinal data (frequency of systemic features) and continuous data (Beighton score and LLAS) were analysed. Spearman's rank correlation coefficient (ρ) of ≤ 0.35 was considered weak, between 0.36 and 0.67 was considered moderate, and ≥ 0.68 was considered strong [30].

Data availability The public release of the dataset of this study was not requested at the time of ethics application. However, all participants of this study have agreed for their data to be used for future studies upon subsequent approval by the University of Sydney Human Research Ethics Committee. Therefore, the datasets generated during and/or analysed during the current study are available from the corresponding author upon reasonable request.

Result

Demographics

A total of 116 participants with a medically confirmed diagnosis of JHS or EDS-HT completed the survey. Fifty-seven participants (53 females) also participated in the physical examination. Participants were predominantly Caucasian and female (Table 1).

Prevalence and frequency of self-perceived systemic features

As the data were not skewed, the prevalence and frequency (mean and standard deviation) of three of the most commonly identified self-perceived systemic features, i.e. orthostatic

Table 1 Summary of demographic data of cohort (n = 116)

Age		Gender		Ethnicity		
Range	Mean (SD)	M	F	Caucasian	Non-Caucasian	Not reported
M 16–53 F 16–68	33.5 (11)	6 (5%)	110 (95%)	108 (94%)	6 (5%)	2 (1%)

M males, *F* females

intolerance, easy bruising, and urinary incontinence (Table 2). Orthostatic intolerance was the most prevalent feature.

Prevalence of generalised joint hypermobility

The mean Beighton and LLAS cutoff scores for the 57 participants who undertook the physical examination were 6.1/9 ± 1.4 and 7.2/12 ± 2.3 respectively. Using the Beighton score cutoffs recommended by Singh and colleagues [27] to identify GJH, 53 of the 57 (93%) tested participants met the cutoff for their respective ages based on gender. The four participants who did not meet the Beighton cutoff for age were females aged 16–39 years who scored < 5/9. Using the LLAS cutoff of ≥ 7/12, 37 participants (65%) were identified as having GJH. Regardless of the Beighton cutoff score used to determine the prevalence of GJH, i.e. ≥ 4, ≥ 5, and age and gender specific, these were significantly different to the cutoff for LLAS at $p < 0.001$ (Table 3).

Correlation between the Beighton score/LLAS and self-perceived systemic features

All correlations between the Beighton score and the self-perceived frequency of easy bruising, urinary incontinence, and orthostatic intolerance were weak and not statistically significant. Similarly, the correlations between the LLAS score and easy bruising, urinary incontinence, and orthostatic intolerance were weak and not statistically significant (Table 4).

The correlations were repeated with the four males removed from the analyses so that the results could be generalised to adult females with JHS/EDS-HT. Despite this, no significant correlations were found in females between the extent of GJH (using either scoring system) and the three systemic features.

Discussion

The findings of this study confirm that orthostatic intolerance, easy bruising, and urinary incontinence are highly prevalent symptoms experienced by people with a medical diagnosis of JHS/EDS-HT.

The prevalence of orthostatic hypotension, in the healthy population, is reported as 5% in people 71 to 74 years increasing with age to 11% at 85 years and over [31], while combined orthostatic hypotension, postural orthostatic tachycardia syndrome, and uncategorised orthostatic intolerance were reported in 10% of healthy individuals of mean age 32 years [23]. Ninety-seven percent of participants in the current study reported experiencing symptoms of orthostatic intolerance. Although females have a higher prevalence of orthostatic intolerance than males [32], the

Table 2 Self-perceived prevalence and frequency of systemic features (n = 116)

Systemic feature	Prevalence (%)	Frequency (%)	Frequency mean (SD)
Orthostatic intolerance	114 (98%)	Normal 1.7% Low 8.6% Moderate 9.5% High 22.4% Very high 57.8%	72.8 (28.0)
Easy bruising	113 (97%)	Normal 2.6% Low 9.5% Moderate 12.1% High 36.2% Very high: 39.7%	65.1 (25.7)
Urinary incontinence	98 (84%)	Normal 15.5% Low 35.3% Moderate 13.8% High 17.2% Very high 18.1%	37.1 (34.4)

Normal = score of 0 mm; low = score 1–25 mm; moderate = score 26–50 mm; high = 51–75 mm; Very high = 76–100 mm for the frequency of each of the symptoms on a 100-mm VAS scale

Table 3 Number of participants who met the cutoff scores for the various classifications of GJH

	Beighton score ≥ 4/9 cutoff	Beighton score ≥ 5/9 cutoff	Beighton score—age- and gender-specific cutoff (Singh et al. 2017)	LLAS
Met the criteria for GJH	56 (98%)	49 (86%)	53 (93%)	37 (65%)

extremely high prevalence found in this study is unlikely to be explained by gender alone. Future research should confirm the prevalence of orthostatic hypotension or postural orthostatic tachycardia syndrome using clinical tests [33].

Quantifying the self-perceived frequency of their orthostatic intolerance, 80% of the participants rated these as high to highly frequent. Orthostatic intolerance is characterised by dizziness, presyncope, and syncope when standing, after exercise, and during/after a hot shower [33, 34]. Moreover, patients with orthostatic intolerance are often unable to maintain prolonged standing posture, requiring a recumbent posture for symptom relief [34]. This greatly limits not only their function in daily activities, such as house chores and participating in recreational activities, but also work that requires prolonged standing posing a risk of injury due to falling. Additionally, orthostatic intolerance is correlated with depression and diminished quality of life [35]. A study of the predictors of the frequency of disabling fatigue experienced by people with JHS/EDS-HT found orthostatic intolerance to be one of four modifiable predictors [12]. However, to date, no studies have investigated the impact of orthostatic intolerance on the health-related quality of life in people with JHS/EDS-HT, and future research is warranted.

Studies of large population samples in Europe, the UK, and the USA confirm that the symptoms of urinary incontinence directly affect health-related quality of life impacting energy, social isolation, and physical mobility, with subsequent impact on mental health [36, 37]. The 84% prevalence of urinary incontinence found in this study is higher than the 69% found in a UK study of 148 adult females with JHS/EDS-HT of a similar age [24]. A recent Australian national report found urinary incontinence affects about 15% of Australian men and women and 38% of women alone aged between 15 and 69 years [38]. The high prevalence found in our study compared to that of the national norms [38] supports the contention that people with JHS/EDS-HT are more prone to experience urinary incontinence than those without the condition. In addition to the stated prevalence, it is likely that health-related quality of life is adversely affected with 35% of participants reporting high to very high frequency of urinary incontinence. Studies of urinary incontinence in other connective tissue disorders report a similar prevalence to that found in people with JHS/EDS-HT [24] suggesting that the causative mechanism behind urinary incontinence in all these disorders may be similar. It has been proposed that the abnormal synthesis of collagen has led to alteration in collagen content in the musculotendinous pelvic floor, rendering it less able to adequately support the internal organs [21]. Ultimately, the internal organs press on the bladder resulting in urinary incontinence [21, 39]. Another potential cause may be due to the presence of abnormal collagen in the supporting structures of the bladder, as pelvic organ prolapse is strongly associated with joint hypermobility [40]. Further research into the mechanism underlying urinary incontinence in connective tissue disorders including JHS/EDS-HT would assist in targeting this distressing symptom.

This study found that easy bruising is a common systemic feature in people with JHS/EDS-HT. Nearly all participants surveyed (97%) reported easy bruising with 76% rating its occurrence as high to very highly frequent. It has been proposed that the underlying cause of easy bruising is fragile capillaries and surrounding connective tissues, as a result of mechanically impaired collagen [21, 41]. While "easy bruising" is listed as a feature of classic, kyphoscoliotic, and arthrochalasis types of EDS and "excessive bruising" as a feature of the vascular and dermatosparaxis types, the Villefranche nosology does not include any reference to bruising for the hypermobility type [5]. Although easy bruising is unlikely to affect an individual's function, it has more covert consequences. Unrecognised as a sign of EDS-HT, children are sometimes suspected to be victims of physical abuse. The unsightly and tender bruising may result in adults abstaining from participating in activities that they perceive as risky, limiting their social engagement, and possibly affecting their physical and mental well-being [42]. Therefore, it is important for healthcare professionals to recognise easy bruising as a systemic feature of JHS/EDS-HT, in order to provide appropriate advice, prevention, and therapy. These will include

Table 4 Correlation between the frequency of generalised joint hypermobility (as measured by the Beighton score and Lower Limb Assessment Score) and self-reported systemic features (all participants)

	Beighton score	Lower Limb Assessment Score
Orthostatic intolerance	$\rho = -0.21, p = 0.11$	$\rho = -0.23, p = 0.08$
Easy bruising	$\rho = -0.08, p = 0.58$	$\rho = 0.10, p = 0.46$
Urinary incontinence	$\rho = -0.18, p = 0.17$	$\rho = 0.08, p = 0.56$

protective pads and bandages in contact sports, behavioural and psychological support [5], and judicious use of supplementation and perioperative medication [43].

The finding of such high prevalence of orthostatic intolerance, urinary incontinence, and easy bruising warrants further investigation of the diagnostic relevance of these symptoms. Additional work would be required to determine the sensitivity and specificity of each symptom within the diagnostic process.

The Beighton score identified significantly more participants with GJH than the LLAS regardless of whether the ≥ 4, ≥ 5, or age- and gender-specific cutoffs were applied. The Beighton score is an upper limb dominant measure of GJH, while the LLAS is a valid measure of both lower limb specific and generalised hypermobility in the adult Caucasian population [27]. The finding of this study, in agreement of the findings of Clinch et al. (2011) [44], would suggest that the hypermobile joints of the majority of our participants were located in the upper limb. Future research should include an upper limb specific hypermobility measure such as the Upper Limb Hypermobility Assessment Tool to better determine the distribution of joint hypermobility in people with hEDS and HSD [45].

No significant correlations were found between the extent of joint hypermobility as measured with the Beighton score and the self-perceived frequency of the systemic features of orthostatic intolerance, easy bruising, and urinary incontinence. This suggests that the extent of generalised joint hypermobility as determined using the five composite tests is not associated with the frequency of three of the most prevalent systemic features of the condition. The same lack of significant correlation was found between the extent of generalised joint hypermobility as measured with the LLAS and the frequency of these systemic features. It appears that the phenotypical presentation of those with the heritable disorder of connective tissue, JHS/EDS-HT, is not as hypothesised. Despite the common collagen types I and III affected, the extent of joint signs and symptoms is unrelated to the frequency of experienced vascular, urogenital, and integumentary symptoms. This suggests that the expression of the collagen deficiency can be organ specific or that the factors affecting these systems are multi-factorial (e.g. physical and mental fatigue, musculature capacity/training, balance and coordination ability). Therefore, it cannot be assumed that an individual with a high degree of generalised joint hypermobility, as assessed using either measure as a continuous variable, will experience more frequent systemic features of syndromic hypermobility.

Strengths, limitations, and future research

A major strength of this paper is the sample size. The study incepted more than 100 participants who were medically diagnosed with JHS/EDS-HT, a sample size which is large for a study of its type. Few studies have a similar sample size. In addition, our study is novel in that it statistically correlated the extent of hypermobility (measured by two validated scoring systems) with the frequency of three commonly reported systemic features of the syndrome. The presence and frequency of systemic features were self-reported. Although this may introduce a degree of bias, symptoms can only be recorded, both clinically and in research, through self-report. Diagnostic accuracy would be improved by employing objective outcome measures such as urinary flow studies and tilt-table tests for orthostatic intolerance. This, however, was not the intention of this study, which was to determine the patient's perception of their symptoms. Another limitation of this study is its generalisability. The study incepted a predominantly female Caucasian population, so its results cannot be inferred to a more culturally or gender diverse population of people with JHS/EDS-HT. Finally, data on physical activity levels were not collected in this study limiting the ability to adjust for physical activity in the analysis. Future studies should aim to recruit people from other ethnicities and utilise multicentred international recruitment strategies to further increase sample size. Further studies should also determine the sensitivity and specificity of the three systemic features surveyed to determine whether they warrant inclusion in future revisions of the classification of hypermobility Ehlers-Danlos syndrome and hypermobility spectrum disorder.

Conclusions

The high prevalence of systemic features, such as orthostatic intolerance, easy bruising, and urinary incontinence in the adult population with JHS/EDS-HT, demonstrates that syndromic hypermobility is not limited to musculoskeletal expressions of the disorder. Healthcare providers should be aware that the extent of musculoskeletal features, notably joint hypermobility, is not indicative of the extent of non-musculoskeletal systemic features in particular orthostatic intolerance, urinary incontinence, and easy bruising. Healthcare professionals should also be aware of the common systemic features when suspecting syndromic hypermobility in order to provide appropriate advice and tailor treatment strategies.

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Compliance with ethical standards

Ethical approval was granted by the University of Sydney's Human Research Ethics Committee (No. 2012/5580).

Disclosures None.

APPENDIX 1

Table 5 The Beighton Score [24]

Test	Score Allocation
1) Passive dorsiflexion of the little fingers beyond 90°	Right: 1 Left: 1
2) Passive apposition of the thumb to the ventral aspect of the forearm	Right: 1 Left: 1
3) Hyperextension of the elbow joint beyond 10°	Right: 1 Left: 1
4) Hyperextension of the knee joint beyond 10°	Right: 1 Left: 1
5) Placing the palms of the hands flat on the floor while maintaining the knees in full extension	1
	Total: score out of 9

APPENDIX 2

Table 6 The Lower Limb Assessment Score [25]

	LEFT		RIGHT	
HIP FLEXION The patient lies supine; the examiner flexes one hip fully; the other leg must stay fully extended on the couch. Does the mid-anterior area of the thigh drop easily onto the stomach/chest with a loose feel to the movement, using a minimum to moderate application of force?	YES	NO	YES	NO
HIP ABDUCTION The patient lies supine, with hip and knees flexed; the knees are dropped outwards and down to the couch, the soles of the feet remain together. With the examiner's hand against the lateral femoral condyle, can the knees come down to the couch sufficiently to let the back of the examiner's hand touch the couch?	YES	NO	YES	NO
KNEE HYPEREXTENSION The patient lies supine; the knees are relaxed and straight, with minimal force, keeping the femoral condyles on the couch, can the heel be lifted at least 3cm off the couch (greater than 2 finger widths)?	YES	NO	YES	NO
KNEE ANTERIOR DRAWER TEST The patient is supine; the hips and knees (90°) are flexed; the examiner gently sits on the foot to stabilise it; moderate pressure is placed against the femoral condyles as the tibia is pulled forwards. Is there a definite, obvious forward movement of the tibia against the femur? Palpable "clunking" of the joint surfaces moving against each is indicative of a positive draw sign.	YES	NO	YES	NO
KNEE ROTATION The patient lies supine; the examiner flexes the hip and knee to 90° and palpates the tibial tubercle; holding the malleoli and ankle firmly, the tibia is rotated medially and laterally on the femur. Normal movement is 1cm medially and laterally. Does the tubercle move easily beyond 1cm in any direction or greater than 2cm overall? With increased internal movement the head of the fibula/lateral condyle of the tibia may also be seen to move.	YES	NO	YES	NO
ANKLE JOINT DORSIFLEXION The patient lies supine; the knee is flexed to 45°; with moderate to strong force the ankle is dorsiflexed. Does the ankle flex more than 15 degrees? Along with the increased movement there may be bulging of the skin and subcutaneous fat anterior to the ankle.	YES	NO	YES	NO
ANKLE ANTERIOR DRAW TEST The patient lies supine; the knee is flexed to 45°; the examiner grasps the heel along the plantar and posterior surfaces with one hand and applies a stabilising force against the anterior of the tibia with the other hand. Using a strong anterior force, can the calcaneum and talus be brought forwards on the tibia? Any forward movement felt is a positive result.	YES	NO	YES	NO
SUBTALAR JOINT INVERSION The patient is supine with their feet over the end of the couch; the examiner holds the posterior surface of the heel and moves the heel into inversion without moving the leg. Is excessive inversion of the subtalar joint seen using minimal force? The sole of the foot or visualisation of the neck of the talus should show movement of 45° inwards, the lateral head of the talus will be very prominent.	YES	NO	YES	NO
MIDTARSAL JOINT INVERSION The patient is supine with their feet over the end of the couch; the midtarsal joint is isolated from the subtalar joint; the forefoot is grasped from lateral to medial along the metatarsals; only minimal-moderate force is applied to invert the midtarsal joint. Does the midtarsal joint invert beyond 45° so that the plantar surface of the metatarsal heads can be brought inwards by 45 degrees?	YES	NO	YES	NO

Table 6 (continued)

	LEFT		RIGHT	
MIDTARSAL JOINT AB/ADDUCTION AND DORSI/PLANTARFLEXION The patient is supine with their feet over the end of the couch; the examiner grasps and stabilises the rearfoot; the forefoot is moved in the direction of ab/adduction and dorsi/plantarflexion. Normal movement should be 1cm in each direction. With minimal force, does the forefoot move easily, almost "wobbling", in an increased amount? Excessive movement in either of the two planes is a positive result.	YES	NO	YES	NO
METATARSOPHALANGEAL MOVEMENT The patient is supine with their feet over the end of the couch; the hallux is dorsiflexed using minimal-moderate force.	YES	NO	YES	NO
Does the hallux dorsiflex easily beyond 90° relative to the metatarsal?				
EXCESSIVE SUBTALAR MOVEMENT The patient is to march on the spot and stop on command; the patient is asked to invert their foot and hold the position close to subtalar joint neutral; the patient is then asked to relax their foot; the movement is observed.	YES	NO	YES	NO
Does the arch lower and flatten fully, excessively and easily, with the talus bulging medially? The pronation noted should be at the end of range of the subtalar joint motion so that no further pronation is possible.				
To score, each limb is calculated separately giving a left score and right score. Each YES is given one mark. A total score of 12 marks is available for each limb, with a score out of 24. The mark is then averaged for a total score out of 12	Total: score out of 12			

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