Arthrometer Assessment of Joint Laxity in People with Ehlers-Danlos Syndrome

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Keywords:	Connective tissue disorder, Ehlers-Danlos Syndrome, Hypermobility, Arthrometer, Joint laxity
Abstract:	Abstract Background: Joint hypermobility is a condition in which synovial joints move beyond normal limits. In children, 10% to 25% experience hypermobility syndrome. Adult hypermobility is reported to range from 5% to 25% in the USA. Joint mobility syndrome includes inherited connective tissue disorders such as Ehlers-Danlos Syndrome (EDS). Typically, a score of 4 or 5 out of 9 on the Beighton scale is indicative of hypermobility in adults. Whereas 6 out of 9 is the criteria for children. No significant correlations were found between the systemic features of EDS and the Beighton score. Purpose: The purpose was to identify clinical techniques/data to contribute to the identification of connective tissue disorders. Methods: A Mobil-Aider arthrometer was used to quantify the anterior and inferior translation of the glenohumeral joint, as well as the anterior translation of the talocrural joint. Results: Thirteen control participants without EDS and 14 participants diagnosed with EDS participated. Significant between-group differences and medium to large effect sizes were found for all 3 motions. Conclusions: The Beighton score has known limitations as diagnostic criteria for hypermobility syndrome and EDS. Testing with an arthrometer provides objective data and can provide a magnitude of hypermobility, not just dichotomous criteria. Clinical Significance: Identification of techniques to obtain objective clinical data are important in the prompt and accurate identification of pathology.

Arthrometer Assessment of Joint Laxity in People with Ehlers-Danlos Syndrome

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Introduction

Clinicians frequently strive to restore joint mobility and function. Yet, manipulations and mobilizations may not be appropriate for all patents.[1] Individuals with hypermobile joints require a different approach. It is important to distinguish the patient who is trained for muscular flexibility from those with generalized articular instability. The value of this differential diagnosis cannot be understated.[2] Systemic joint hypermobility is a chronic condition and requires lifelong support.[2]

Joint hypermobility is a condition in which synovial joints move beyond normal limits.[3] Estimates of the frequency of hypermobility syndrome are significant. In children, 10% to 25% experience hypermobility syndrome.[4,5] Adult hypermobility is reported to range from 5% to 25% in the USA, 25% to 38% in Iraq, and 43% in the Noruba tribe in Nigeria.[6-10] Cooper and Brems found 76% of surgical patients with multi-directional glenohumeral instability demonstrated generalized joint hypermobility.[11]

Joint mobility syndrome includes inherited connective tissue disorders such as Ehlers-Danlos Syndrome (EDS).[12] EDS affects many systems of the body.[13-22] The 2017 International Classification recognizes 13 subtypes of EDS.[23] The Villefrache subtypes include: classical, hypermobility, vascular, kyphoscoliosis, arthrochalasia, and dermatosparaxis.[24] The hypermobility type (hEDS) is the most common and represents 80% to 90% of EDS cases.[23,25] Individuals with EDS often have poor muscle definition and adopt end-range postures.[3] A typical standing posture may include flat feet, hyperextended hips and knees, increased lumbar lordosis, and "hip hanging,"[3] Clinical diagnostic criteria have included the Beighton Scale (figure 1) and Brighton Criteria (figure 2). However, the diagnosis of joint hypermobility should also include examination of skin elasticity, scars (thin), stretch marks (adolescent growth spurts), hernia, pelvic floor, varicose veins, Gorland's sign (tip of the tongue to the nose), and the absence of a frenulum. While some of these items do appear in the second criterion of the diagnostic criteria from 2017, a formal diagnosis cannot be made at this time if the Beighton Scale requirement is not met. Typically, a score of 4 or 5 out of 9 on the Beighton scale is indicative of hypermobility in adults, whereas 6 out of 9 is the criteria for children. No significant correlations were found between the systemic features of EDS and the Beighton score.[26] Furthermore, the Beighton Score does not differentiate between congenital articular instability versus trained hypermobility. Factors that influence the Beighton Score may include:

1. A patient with EDS may not demonstrate a "positive" score because of muscular guarding/tightening as a protective factor (e.g.: hamstrings in palms to floor test).
2. Individual anatomy may limit people with true connective tissue disorders in instances such as bony end feel (elbow extension or knee hyperextension).
3. People who may have trained for enhanced muscular flexibility (dancers, gymnasts) and do not necessarily have joint instability. Thus, they may score high on this test without the dangers of subluxation or dislocation.
4. The test currently examines a series of joints that are not most typical of dislocations/subluxations. The Beighton Score does not address the shoulders, hips, or ankles (most problematic lax joints).

Thus, the purpose of this study was to objectively quantify joint laxity of the shoulders and ankles in a control group and that of a group known to be diagnosed with EDS. The joint laxity was quantified with an arthrometer to compare the two groups as well as the magnitude of joint laxity compared to the Beighton Score.

Methodology

Level of Evidence II. The consent form, approved by the Institutional Review Board for the Protection of Human Subjects (#87-22) was reviewed and signed by the potential participant. Each person was screened for inclusion criteria. All participants were over 18 years of age. All participants were assessed with the Beighton Scale. The testing researcher was blinded to the Beighton Scale score. Participants in the control group were required to have a "zero" score and no injury or surgery to the shoulder or ankle. Participants in the EDS group were expected to have a high Beighton score but shoulder or ankle joints with a current injury or prior surgery were eliminated from data collection. Thus, both shoulders and ankles were tested on some people but not all. Demographic data included age and gender.

The device used in this study was the Mobil-Aider arthrometer (figure 3). This arthrometer has a stable side (red side with LED screen) and a side that moves linearly (black side without screen) via an internal rollerball mechanism. Each side of the main body of the device accommodates contoured attachments for a variety of joints. In this study ankles and shoulders were tested. For the ankle, the yellow convex attachment contours to the posterior distal tibia (gastroc/soleus/Achilles region) while the black concave attachment conforms to the talus/calcaneal region. Both pieces were locked into position on their respective sides of the device via a dovetail fit and plugger mechanism. The axis of the Mobil-Aider was aligned with the talocrural joint line. The proximal side (yellow) of the Mobil-Aider was stabilized against the posterior tibia. The distal side (black) of the Mobil-Aider was held in contact with the talus/calcaneus. For the shoulder, an inferior translation was performed with the green contoured attachment on the proximal side and the blue attachment was used for anterior translation.

Participants were positioned comfortably for the three testing procedures.

- Shoulder inferior translation = supine with arm relaxed at their side, hand on the belly with forearm pronated, and a towel roll under the elbow.
- Shoulder anterior translation = prone with the arm at their side and a small wedge under the ipsilateral clavicle/anterior chest
- Ankle anterior translation = prone with feet over the edge of the table and a small wedge placed under the distal lower leg

The axis of motion of each joint was identified with the passive range of motion performed by the researcher. The Mobil-Aider arthrometer axis was aligned with the joint line. The proximal element of the Mobil-Aider was stabilized against the proximal bone as follows:

- Shoulder inferior translation = stabilize upper thorax/upper chest (figure 4)
- Shoulder anterior translation = stabilize scapula (figure 5)
- Ankle anterior translation = stabilize tibia (figure 6)

The distal segment was mobilized as follows:

- Shoulder inferior translation = apply a distal force through the humeral head/shoulder bone (figure 4)

- Shoulder anterior translation = apply an anterior force to the posterior humeral head/shoulder bone (figure 5)
- Ankle anterior translation = apply an anterior force through the talus/calcaneus/back of foot (figure 6)

A few small amplitude test oscillations were performed to confirm proper positioning. Then three movements of each motion (shoulder inferior translation, shoulder anterior translation, ankle anterior translation) were performed with a 30-second rest between tests. Each data point was recorded. Measures were reported in millimeters of linear translation. After the testing of each individual, the surfaces of the Mobil-Aider™ and wedges were cleaned with anti-microbial wipes.

Results:

Thirteen control participants without EDS and 14 participants diagnosed with EDS participated in the study. In the control group, 6 participants were male and 7 were female. In the EDS group, 1 participant was male, and 13 were female. The mean age of the control group was 24.1 (± 3.4) and of the EDS group was 32.4 (± 12.1). In cases where the bilateral shoulder or ankle joint met inclusion criteria, these measurements were recorded as a separate case. Thus, for shoulder anterior translation there were 23 in the control and 26 in the EDS group. For shoulder inferior translation there were 21 in the control and 26 in the EDS group. Finally, for the ankle anterior translation, there were 22 in the control and 25 in the EDS group. An independent samples t-test demonstrated a significant between-group difference for age ($p = .026$). The control group was required to have a Beighton score of 0; the EDS group was required to have a Beighton score greater than 6. The EDS group had a mean Beighton score of 8.0 (± 1.2).

The average of 3 trials was taken for each motion and then Mann-Whitney U tests were performed to identify between-group differences for all three joint translations measured: anterior and inferior shoulder glide and anterior ankle glide. Effect sizes were calculated using Cohen's d formula: Cohen's $d = M_1 - M_2 / s_{pooled}$ where $s_{pooled} = \sqrt{[(s_1^2 + s_2^2) / 2]}$.[27] Effect size r_{Yl} was then calculated using the formula $r_{Yl} = d / \sqrt{(d^2 + 4)}$. Significant between-group differences and medium to large effect sizes were found for all 3 motions (table 1).

A priori power analysis concluded that 42 total participants would be needed given an assumed effect size of 0.8, the desired power of 0.8, and an alpha level set at 0.05.[28,29] The post-hoc analysis affirmed that the study was sufficiently powered with 99% power for all data.

Discussion

Joint hypermobility is a topic of interest in the arts, sports, and medical communities.[30] However, the lack of awareness of hypermobility syndrome among healthcare providers can lead to significant delays in gaining a diagnosis.[31] Individuals are told the problems are "growing pains," "all in your head," or they are "malingerers."[31] Some individuals have reported they feel their healthcare provider is dismissive or has "given up" on them.[31] Furthermore, when an individual has hypermobility syndrome, they may be conflicted on whether to participate in sports activities or protect themselves from injury. This can be particularly problematic for parents of children with hypermobility syndrome.

To date, the Beighton scoring system is the most common tool used for the identification of generalized joint hypermobility (GJH). When it was developed in 1973, it was proposed as an epidemiological screening tool, not a clinical tool.[32] The Beighton Score is one of the two major components of the Brighton Criteria and is used for the diagnosis of joint hypermobility syndrome and the hypermobility type of EDS.[33] However, despite numerous studies, the cut-offs that differentiate individuals with and without GJH have not been well defined. The range in the literature is from >4 to >8.[34,35] When using a Beighton cut-off score of >4 for the entire population, a high false-positive rate of 60% occurred, suggesting an overestimation of prevalence.[30] Singh et al (2017) studied 1000 individuals from 3-101 years of age.[32] A logistic regression indicated a false-positive rate of 60.0% and a false-negative rate of 12.4%, with the Beighton scoring system having a sensitivity of 0.8% and a specificity of 99.3% if a cut-off of >4 was used to determine GJH. Based on the Australian cohort for females are suggested the following Beighton scores for GJH:

- >6 for females & >5 for males aged 3-7 years
- >5 for females & >4 for males aged 8-39 years
- >4 for females & >2 for males aged 40-59 years
- >3 for females aged 60-69 years; >1 for males 60+ years
- >2 for females aged 70+ years

Thus, a single cut-off score does not appear to be appropriate. In addition, the Singh et al[32] study did not address ethnic differences. The Beighton system also samples a limited number of joints in a single plane of motion. Commonly lax joints which as shoulders, hips, and ankles are not assessed. The purpose of this study was to demonstrate the ability to quantify the magnitude of joint laxity instead of a dichotomous (all-or-nothing) presentation. Technology is available to assist clinicians with the quantification of joint laxity. This study used a Mobil-Aider arthrometer to demonstrate the ability to test multiple joints (ankle in 1 plane & shoulder in 2 planes) and revealed a statistically significant difference between the individuals with and without high Beighton Scores. The mean joint translation of the EDS group was close to double that of the control group (table 1).

In conclusion, testing with an arthrometer has the potential to yield results across multiple joints in different planes to substantiate the diagnosis of GJH. Given the recent availability of a joint arthrometer to test joints other than the knee (KT1000), it will take time to populate the data with normative values across multiple joints. Recent arthrometer publications related to knee laxity, ankle sprains, shoulder comparisons to electromagnetic devices, and wrist inter/intra-rater reliability are steps in that direction.[36-40] Objective data enhances our ability to make clinical decisions and the use of an arthrometer can contribute. Future work needs to continue to expand this database in both normal and conditions of pathology.

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Ethics and consent: The consent form, approved by the Institutional Review Board for the Protection of Human Subjects (#87-22) was reviewed and signed by each participant.

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Conflict of interest: Dr. Dawn T Gulick holds the Mobil-Aider arthrometer patent.

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Figure 1. Beighton Scale

1. right thumb to radius
2. left thumb to radius
3. right 5th digit hyperextension >90 degrees
4. left 5th digit hyperextension >90 degrees
5. right elbow hyperextension >15 degrees
6. left elbow hyperextension >15 degrees
7. right knee hyperextension >15 degrees
8. left knee hyperextension >15 degrees
9. palms touch the floor with legs straight

Figure 2. Brighton Criteria

Major Criteria:
- Beighton score ≥ 4 out of 9
- Arthralgia present in ≥ 4 joints for 3 months

Minor Criteria:
- Beighton score ≤ 3 out of 9
- Arthralgia present in ≤ 3 joints (or back pain) for ≥ 3 months
- Dislocation/Subluxation of ≥ 1 joints, ≥ 1 times
- ≥ 3 soft tissue lesions (bursitis, epicondylitis, tenosynovitis)
- Marfanoid habitus
  - Wingspan to height ratio > 1.03
  - Upper:Lower segment ratio < 0.89
  - (+) Steinberg sign
- Abnormal skin: hyperextensibility, scarring
- Eye signs: eyelids drop, myopia
- Varicose veins; hernia, uterine, or rectal prolapse

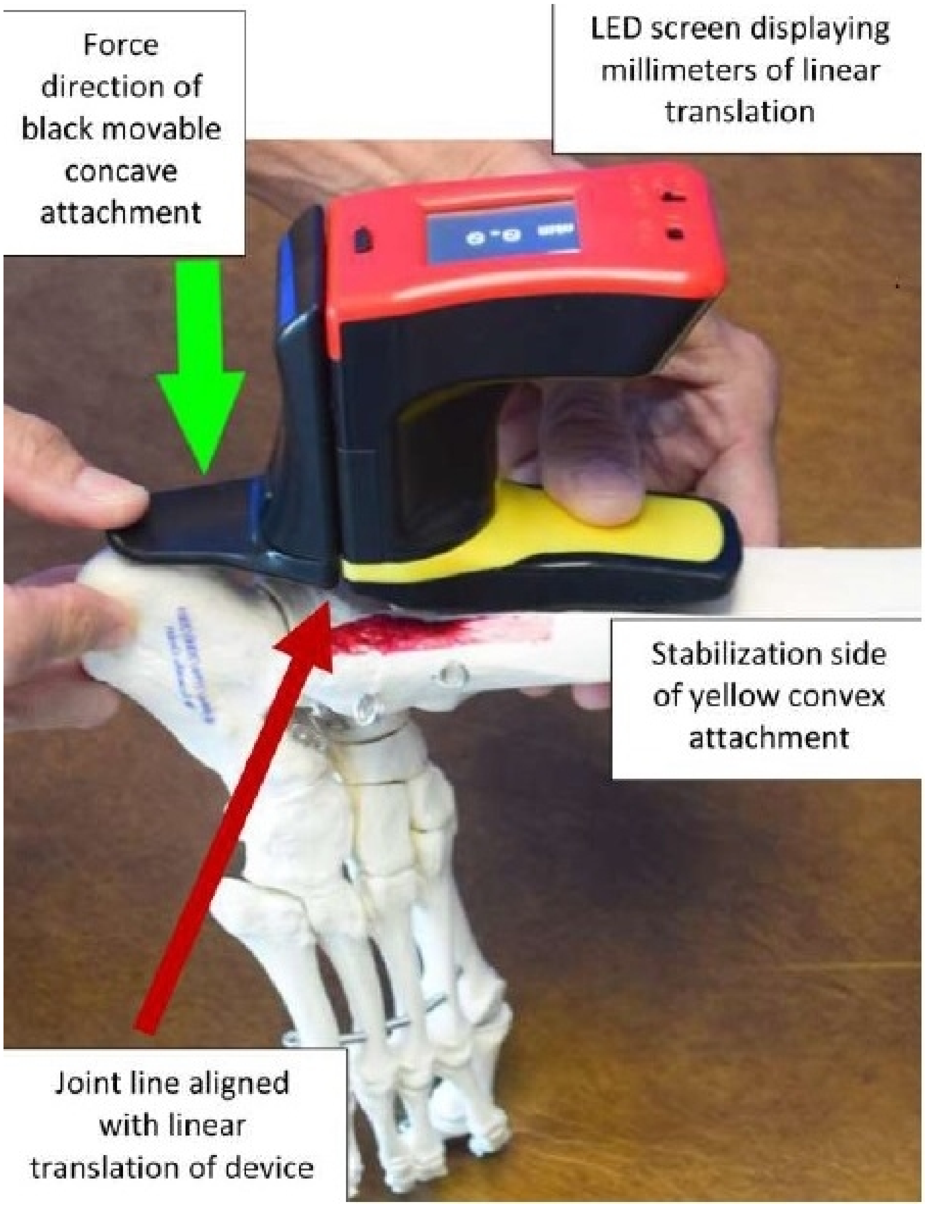

Figure 3. Function of the Mobil-Aider Arthrometer

83x107mm (144 x 144 DPI)

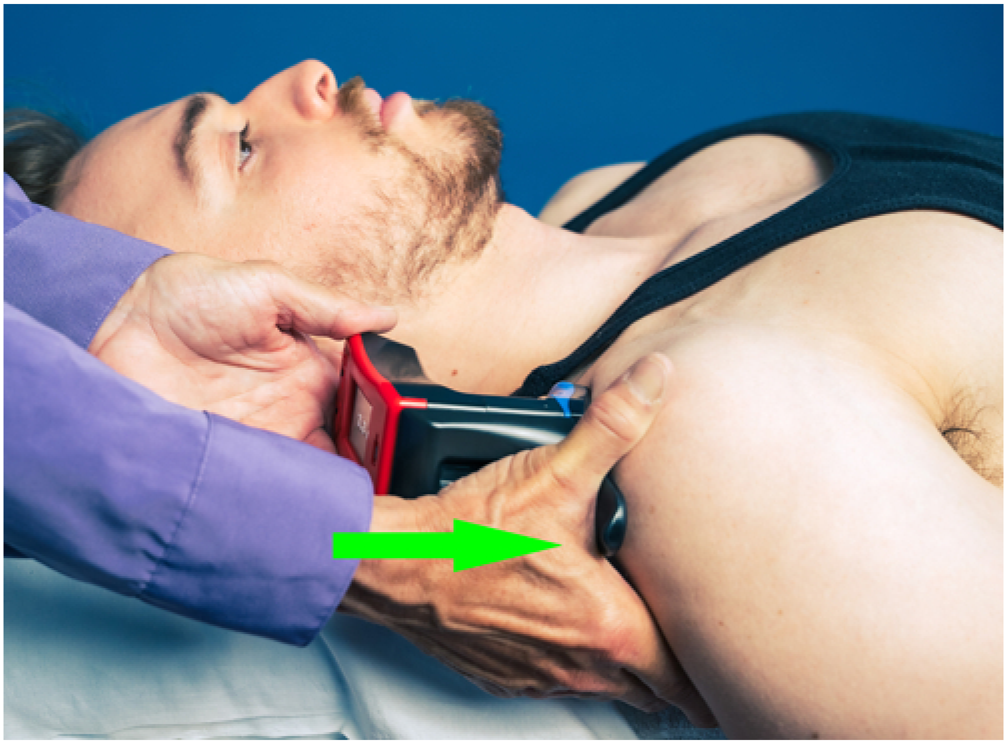

Figure 4. Shoulder Inferior Translation with the Mobil-Aider

107x79mm (118 x 118 DPI)

Figure 5. Shoulder Anterior Translation with the Mobil--Aider

107x90mm (118 x 118 DPI)

Figure 6. Talocrural Anterior Translation with the Mobil-Aider

107x91mm (118 x 118 DPI)

Table 1. Between-group differences of joint laxity as tested with an arthrometer

	Control		EDS			
	Mean	SD	Mean	SD	p-value	Effect size
Anterior shoulder translation	5.45	1.43	10.56	1.74	<.001	.85
Inferior shoulder translation	4.27	1.60	8.51	1.63	<.001	.80
Anterior ankle translation	5.36	1.19	8.07	1.84	<.001	.66