Foot Type Analysis Based on Electronic Pedobarography Data in Individuals with Joint Hypermobility Syndrome/Ehlers-Danlos Syndrome Hypermobility Type During Upright Standing

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Background: Joint hypermobility syndrome/Ehlers-Danlos syndrome hypermobility type (JHS/EDS-HT) is a rheumatologic condition characterized by generalized joint hypermobility and musculoskeletal and nonmusculoskeletal findings related to congenital laxity of connective tissue. Because foot pain and other foot problems are reported to make daily life problematic to manage for individuals with JHS/EDS-HT, and thanks to the availability of modern technology, the aim of the present study was to quantitatively characterize foot type in individuals with JHS/EDS-HT during upright standing.

Methods: Forty feet of 20 women with JHS/EDS-HT (mean ± SD age, 36.03 ± 14.01 years) were assessed clinically and with a pressure-sensitive mat during upright standing.

Results: Forty-five percent of feet had a high arch (pes cavus), 27.5% had a normal arch, and 27.5% had a low arch (pes planus or flatfoot).

Conclusions: From a clinical perspective, the characterization of foot type in JHS/EDS-HT is important to identify, develop, and enhance the rehabilitative options. An understanding of the relationship between pes cavus and foot pain in these patients could, in fact, improve the clinical management of these patients. (J Am Podiatr Med Assoc 104(6): 588-593, 2014)
This condition seems to affect no less than 1 in 10,000 in the general population and is dramatically more common in women. Major features include joint hypermobility, joint complications, and minor skin features (eg, skin hyperextensibility), and the presence of additional cutaneous, vascular, skeletal, and ocular findings moves toward the diagnosis of other EDS variants. In JHS/EDS-HT, problems in the lower limbs, especially in connection with the feet, are common and vary in presentation and severity. The musculoskeletal system is clearly one of the most commonly and severely affected anatomical structures, with hyperextensibility of ligaments, tendons, and capsules. In addition to capsuloligamentous laxity, various studies demonstrated lack of proprioception at the knees, shoulders, and proximal interphalangeal joints. Several authors have reported on flat feet in JHS/EDS-HT, hallux valgus, joint contractures, and other orthopedic problems directly associated with the foot.

Focusing on the characterization of foot type in individuals with JHS/EDS-HT, the scientific literature is extremely limited, out of date, and mainly based on clinical examinations. To our knowledge, the only study was conducted by Beighton and Horan, which assessed 100 patients with JHS/EDS-HT. They found that the most common foot deformity was severe pes planus (52%), which was usually asymptomatic; in some of the younger patients, the longitudinal arch was normal when no weight was being borne, but by age 30 years, all of the patients with affected feet showed static and dynamic planus deformity. Similar results were found by Berglund et al. However, no precise indications about the technique used to assess the presence/absence of flatfoot are presented in these papers.

Plantar foot pressure measurement with a pressure-sensitive mat is helpful in investigating underfoot pressure during standing and during gait. It is a noninvasive method that is valuable for diagnosing foot diseases and defects and for monitoring treatment. It can be used to examine children and adults with a variety of foot impairments associated with neurologic and musculoskeletal disorders. With this method, there is the opportunity to obtain a quantitative static/dynamic analysis of footprints based on the distribution of plantar pressures during contact of the foot with the ground. In this way, it is possible to obtain a readily available and noninvasive method to measure the footprints and the deformation of the foot arch during weightbearing activities.

Because foot pain and other foot problems are reported to make daily life problematic to manage for individuals with JHS/EDS-HT, and thanks to the availability of modern technology, the aim of the present study was to quantitatively characterize foot type in individuals with JHS/EDS-HT during upright standing using a baropodometric platform. In this way, it could be possible to better investigate and increase the knowledge about podiatric medical features in patients with JHS/EDS-HT.

Materials and Methods

Participants

Twenty women with JHS/EDS-HT (mean ± SD: age, 36.03 ± 14.01 years; height, 1.65 ± 0.08 m; and weight, 59.95 ± 12.03 kg) were enrolled in this study, for a total of 40 evaluated lower limbs. The diagnosis of JHS/EDS-HT was established using published Brighton criteria and the Villefranche criteria. Patients were included if they met at least either one of these two sets. Because JHS/EDS-HT is a diagnosis of exclusion, the absence of features suggestive of other heritable connective tissue disorders was assessed in a clinical genetics outpatient clinic.

All of the participants were asked to complete the Fatigue Severity Scale (FSS) and the 11-point Numeric Rating Scale (NRS-11) for pain. The FSS is a questionnaire to evaluate the intensity of fatigue; in particular, it is a scale quantifying fatigue intensity, which has been used in different chronic conditions, showing high internal consistency and validity. The FSS is composed of nine items with a 7-point response format that indicates the degree of agreement with each statement. The score considered is the mean value of the items; the minimum mean score possible is 1 and the maximum is 7. Higher scores represent greater fatigue severity. The NRS-11 for pain is an 11-point numeric scale used to grossly measure every kind of pain with a score ranging from 0 (no pain) to 10 (the worst pain).

The study was approved by the ethics research committee of the Istituto di Ricovero e Cura a Carattere Scientifico (IRCCS) “San Raffaele Pisana,” Tosinvest Sanità, Roma, Italy, and written informed consent was obtained from the patients.

Experimental Setup

We studied patients who could walk without devices, who did not undergo surgery in the 10 years before the evaluation, and who were not
pregnant; all of the patients walked in normal shoes without orthotic devices.

All of the participants were evaluated instrumentally at the Movement Analysis Laboratory of the IRCCS “San Raffaele Pisana,” Tosinvest Sanita`, Rome, Italy. Plantar pressure measurements were performed by means of a pressure-sensitive mat (Tekscan Inc, South Boston, Massachusetts) composed of 2,016 sensing elements arranged in a $42 \times 48$ matrix and connected via USB interface to a personal computer. Participants were asked to stand as still as possible on the mat for 5 sec, barefoot and with the feet placed in a comfortable position at a distance not larger than the pelvis width.

Forty temporal frames (sampled at 8 Hz) were acquired for each trial, and matrices containing the plantar pressure value for each element of the sensitive grid were exported as text files for further analysis.

Data Analysis

To identify foot types, we calculated the arch index (AI) from the plantar pressure according to Cavananagh and Rodgers. First, the foot-ground contact area (toes excluded) was divided into three regions, ie, forefoot (A), midfoot (B), and rearfoot (C). Three relative contact areas were estimated and then used to calculate AI according to the following equation (Fig. 1A):

$$\text{AI} = \frac{B}{A + B + C}$$

To derive these three areas, a foot axis line was drawn from the middle of metatarsals 2 and 3 to the middle of the heel. Perpendicular to this foot axis, the foot excluding the toes was divided in three equal parts. Thus, the AI was essentially a ratio of midfoot area to total foot contact area without the toes (Fig. 1B). Based on the AI, plantar arches were classified as follows: $0.21 \leq \text{AI} < 0.26$, normal arch; and $\text{AI} \geq 0.26$, low arch. This procedure was performed by the same operator to ensure data reproducibility; the whole process was performed by means of a custom Matlab routine (The MathWorks Inc, Natick, Massachusetts) that processes the matrices exported by the platform system.

Statistical Analysis

According to the classification of plantar arches described previously herein, the EDS group was divided into three subgroups: high arch ($\text{AI} \leq 0.21$), normal arch ($0.21 < \text{AI} < 0.26$), and low arch ($\text{AI} \geq 0.26$). Kolmogorov-Smirnov tests were used to verify whether the anthropometric measures were normally distributed; the parameters were not normally distributed, so we used nonparametric analysis. Data from the three subgroups were compared using the Kruskal-Wallis test, followed by post hoc comparison, to detect significant differences. Null hypotheses were rejected when $P < .05$.

Results

In the considered group of patients, we found the following distribution: the group with high arch (pes cavus) ($\text{AI} \leq 0.21$) comprised 18 feet (45.0% of evaluated feet), the group with normal arch ($0.21 < \text{AI} < 0.26$) comprised 11 feet (27.5% of evaluated feet), and the group with low arch (pes planus or flatfoot) ($\text{AI} \geq 0.26$) comprised 11 feet (27.5% of evaluated feet).

Eleven participants presented a specific foot type bilaterally; regarding the other participants, one showed foot cavus and flatfoot, four presented flatfoot and normal foot, and four exhibited foot cavus and normal foot.

Regarding clinical evaluations, the FSS mean ± SD score in this population showed high values (5.6 ± 1.9; maximum mean value is 7, indicative of great fatigue severity), evidenced a high fatigue level of the analyzed patients, and the NRS-11 score ranged from 6 to 10, without any differences among the three subgroups. In addition, no significant differences were found among the three subgroups in terms of age or anthropometric measures (height and weight).

Discussion

The JHS/EDS-HT is recognized as a severely disabling condition, with chronic pain, fatigue, and movement and postural alterations as major determinants of severe deterioration of quality of life. Individuals with JHS/EDS-HT endure difficulties with their mobility due to their foot problems and related disability. It is, therefore, of extreme importance to find ways to assess the actual foot status for this group and to support individuals with solutions to their problems with ambulation. In light of these considerations, we assessed plantar arch types in a group of patients with JHS/EDS-HT in static upright position using a pressure-sensitive mat according to a widely vali-
dated method. According to the present data, we observed that most evaluated participants (45.0% of feet) exhibited pes cavus, which has not yet been documented by the literature.

Pes cavus is a term used to describe a foot type with an excessively high medial longitudinal arch. This foot type occurs bilaterally in 8% to 10% of the population. Individuals with this deformity often complain of difficulty with footwear and painful callus formation beneath the metatarsal heads. Rearfoot varus and compensated forefoot valgus predispose patients to lateral ankle trauma and recurrent sprains, resulting in chronic ankle pain as the chief complaint. Almost two-thirds of the participants with pes cavus reported current musculoskeletal foot pain compared with just 23% of individuals with a normal foot type. It is demonstrated that pes cavus is more susceptible than the normal foot to painful conditions in the foot. In addition, pes cavus is characterized by abnormally high pressure-time integrals, which are significantly related to foot pain.

For this reason, it is difficult to compare rigorously the present results with previous ones.

The presence of a high incidence of pes cavus during upright standing that we found in patients with JHS/EDS-HT could be explained as a strategy to better maintain balance, which is generally impaired in this pathologic disorder. It is demonstrated, in fact, that individuals with EDS display decreased postural control mainly due to reduced muscle strength, joint/extremity pain, fatigue, and impaired proprioception. Ligament laxity and muscle weakness, the main features of JHS/EDS-HT, are strictly connected with the presence of flexible flatfoot, particularly present in childhood, which is characterized by the absence of a medial arch over the weightbearing surface but is not a real flatfoot. The flexible flatfoot may disappear with growth, leaving different strategies of foot strike. The presence of pes cavus may also be explained by the presence of an abnormal digit position defined as claw toe due to a muscle hypotonia, in particular of interossei and lumbrical muscles. This strategy is defined as flexor stabilization and typically occurs with the prevalence of flexor digitorum longus activation. As concerns the clinical evaluations, we found no differences in terms of fatigue or pain according to the different foot types, as evidenced by FSS and NRS-11 scores. These data showed that patients with JHS/EDS-HT present with a high fatigue level and the presence of pain. These elements, together with the mentioned foot deformities, may restrict activities at home or

Figure 1. A, Calculation of the arch index (AI). The length of the footprint excluding the toes (L) is divided into equal thirds. The AI is then calculated as the area of the middle third of the footprint divided by the entire footprint area \( \text{AI} = \frac{B}{A + B + C} \). B, The visual AI categorization tool.
at work during standing, walking, or other activities, limiting their quality of life.30-33

From a clinical perspective, precise and quantitative characterization of foot type in JHS/EDS-HT is important to identify, develop, and enhance the rehabilitative options. Untreated pes cavus can be disabling and can result in significant difficulties for the patient and clinicians. An understanding of the relationship between pes cavus and foot pain in these patients will improve their clinical management. For example, the use of foot orthoses could reduce plantar pressure and decrease foot pain in individuals with JHS/EDS-HT. For this reason, further studies should be conducted integrating the results obtained in this study, which are related to characterization of foot type in a static condition, with the foot characterization during gait. In this way, it could be possible to understand whether the presence of pes cavus in patients with JHS/EDS-HT is a peculiarity only of the upright position or whether it is present also during a dynamic task such as gait.

A possible bias of this study is the relatively small sample size, resulting in limited strength of these findings. However, to our knowledge, it represents the first attempt to quantify foot type in patients with JHS/EDS-HT during standing using a validated and rigorous method. In addition, note that JHS/EDS-HT is a relatively rare condition and that large experimental groups are difficult to gather. Further studies should be conducted to confirm these data considering larger groups of patients and comparing this classification approach with other methods and against a clinical or radiographic gold standard used to defined arch height. In addition, how the different foot types could influence balance maintenance during standing and the gait strategy in JHS/EDS-HT may be topics of further investigations. A better understanding of the relationship between abnormalities of the foot and alteration in posture and lower-extremity kinematics is, in fact, important in these patients because these elements could predispose an individual to additional musculoskeletal injuries. In addition, the integration of instrumental data with other clinical assessments could be conducted in the future; at this time, only the FSS and the NRS-11 have been included in the patient assessment, but other clinical evaluations, such as the Manchester Foot Pain and Disability Index, could provide additional information on this topic.

**References**


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**Conflict of Interest:** None reported.