Are patients with hereditary spastic paraplegia different from patients with spastic diplegia during walking? Gait evaluation using 3D gait analysis

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Summary

Patients with hereditary spastic paraplegia (HSP) often resemble patients with mild spastic diplegia (SD), although their motor limitations differ. The aim of this study was to analyse quantitatively the gait of HSP and SD subjects in order to define the gait pattern in HSP and the differences between the two conditions. Fifteen subjects with HSP, 40 patients with SD and 20 healthy subjects underwent gait analysis (GA). The spatio-temporal and kinematic parameters at the proximal joints were found to be similar in HSP and SD, whereas the most significant differences were found at the knee and ankle joints. Both groups displayed a tendency for knee hyperextension in the midstance phase, but the duration of this hyperextension was longer in the HSP patients. This study shows that GA complements traditional clinical evaluations, making it possible to distinguish, clearly, between motor ability in HSP and in SD patients; the duration of the knee hyperextension during midstance was found to discriminate between the two gait patterns.

KEY WORDS: gait analysis, hereditary spastic paraplegia, kinematics, kinetics, rehabilitation, spastic diplegia.

Introduction

Hereditary spastic paraplegias (HSPs) are neurodegenerative disorders characterised mainly by progressive lower limb spasticity. They are classified as “pure” when the spastic paraplegia exists in isolation, and as “complicated” when other major clinical features are also present. Their reported incidence is between 2.0 and 9.6/100000 (1,2). The features whose presence determines this differentiation between the pure and complicated forms include neuropathy, ataxia, retinal pigmented degeneration, optic atrophy, deafness, and mental retardation. The pure forms may also include upper limb hyperreflexia, impaired lower limb vibration sense, and bladder disturbances. HSP may be inherited as autosomal dominant, recessive and X-linked forms (3,4).

As regards the main clinical features of HSP, most patients present difficulty walking or gait disturbances, namely spasticity, hyperreflexia, and extensor plantar responses, with weakness of a pyramidal distribution in the lower limbs (4). In those with childhood onset, a delay in walking is not uncommon. In particular, children with HSP often resemble patients presenting with a mild form of spastic diplegia secondary to cerebral palsy (CP) (5). Up to 25% of affected patients are asymptomatic, and patients who have only subclinical manifestations can escape diagnosis, hence the importance of careful clinical evaluation.

In the literature, the analysis of gait patterns in HSP patients has been limited to clinical examination and a few studies that have quantified gait strategy in adults with HSP, using 3D gait analysis (GA) (2,6). These studies quantified gait pattern in HSP on the basis of spatio-temporal parameters and some kinematic indices of the main lower limb joints; kinetic evaluations were not performed. On the other hand, to the best of our knowledge, analyses of biomechanical strategy during gait in children with HSP are lacking in the literature.

From a clinical viewpoint there is thus a need to investigate, in depth, the gait patterns of paediatric patients with HSP: quantitative analysis of walking, in fact, complements traditional clinical evaluations, yielding information crucial in establishing the level of the functional impairment and identifying the most appropriate therapeutic programme for the patient.

Thus, since children with HSP often resemble children affected by a mild form of spastic diplegia secondary to CP, this evaluation could be an important tool, helping to establish accurately both the quantitative differences between HSP and SD gait strategy and specific treatment programmes for the two pathologies.

The main objective of this study was to identify and quantify the functional limitations (reflected in their gait patterns) of a group of patients affected by HSP and of another group with mild SD secondary to CP, in order to define quantitatively the differences between the two groups, using kinematic and kinetic parameters derived from GA data.
Materials and methods

Subjects

Fifteen patients with a clinical diagnosis of HSP (age range: 6-15 years) and 40 patients with SD secondary to CP (age range: 4-17 years) were evaluated. The selection criteria for the patients with HSP were: a clinical diagnosis of spastic paraplegia in the absence of structural/spinal cord/cerebral disorders, demyelinating, metabolic or inflammatory disorders (particularly for the sporadic forms) and, for the familial forms, the presence of a positive family history of gait disturbances and/or gene mutations (most commonly in the SPG4 and SPG7 genes). The selection criteria for the patients with SD were a physician’s diagnosis of SD, classified as type III according to Rodda’s classification (7), and mild spasticity of the lower limb joints, no history of cardiovascular disease, and no previous surgery or other significant treatments for spasticity. All the patients were able to walk independently without the use of crutches, walkers or braces.

A control group of 20 healthy subjects (Control Group: CG; age range: 5-16 years) was included. The selection criteria for these non-disabled subjects included no history of cardiovascular, neurological or musculoskeletal disorders. The members of the CG exhibited normal range of motion and muscle strength, and had no apparent gait abnormalities. The demographic data of the patients and the healthy subjects are reported in Table I. All the subjects were volunteers and their parents gave their informed consent to the children’s participation in the study. This study was approved by the ethics committee of the IRCCS Eugenio Medea, “La Nostra Famiglia” Association in Bosisio Parini (Italian province of Lecco), Italy and by that of the IRCCS “San Raffaele” Hospital – Tosinvest Sanità Group in Rome, Italy.

The subjects were evaluated in the motion analysis laboratories of the IRCCS Eugenio Medea – “La Nostra Famiglia” Association in Bosisio Parini, or the IRCCS “San Raffaele” – Tosinvest Sanità Group in Rome. The complete evaluation consisted of three parts: clinical examination, videotaping and 3D GA. In the clinical examination, the severity of the spasticity was assessed using a modified Ashworth scale of muscle spasticity (ASH) (8); the ASH was calculated as the mean value of spasticity in both the lower limbs. The child’s overall functional abilities were evaluated using the Gross Motor Functional Measure (GMFM).

The GA was conducted using an optoelectronic system with passive markers (ELITE 2002, BTS, Milan, Italy) working at a sampling rate of 100 Hz (for the kinematic movement evaluation) (9), two force platforms (Kistler, CH) (for the calculation of the movement kinetics), and a video system synchronised with the optoelectronic and force platform systems (BTS, Milan, Italy). After collection of some anthropometric data (height, weight, tibial length, distance between the femoral condyles or diameter of the knee, distance between the malleoli or diameter of the ankle, distance between the anterior iliac spines), passive markers were placed at special points of reference, directly on the subject’s skin, as described by Davis (10).

Each subject was asked to walk barefoot at their own natural pace (self-selected speed) along a 10-metre walkway containing two force platforms at the mid-point. He or she was asked to start from a pre-defined point so as to ensure placement of just one foot on each of the force platforms. From the three-dimensional coordinates of the markers, measured by the optoelectronic system, the angles of main lower limb joints were calculated using Eulerian angles (10). All the graphs obtained were normalised as a % of the gait cycle. Five trials were collected for each child in order to guarantee the consistency of the results. In order to quantify differences between the gait strategies of the two pathological groups and their deviations from normality, several indices from kinematic and kinetic data were identified and calculated: spatio-temporal parameters, joint angle values (pelvis, hip, knee and ankle joints) at specific instants of the gait cycle and in the sagittal plane of movement, and the ankle joint power plot. In detail, the outcome parameters used were the following:

Spatial-temporal parameters:

- % Stance: % of gait cycle that begins with initial contact and ends at toe-off of the same limb;
- Velocity: mean velocity of progression (m/s);
- Step width: medio-lateral distance between the two feet during double support (mm);
- Anterior step length: longitudinal distance from one foot strike to the next one, normalised to subject’s height.

Kinematics:

Pelvic tilt
- Mean Pelvic Tilt: mean value of pelvic tilt angle (degrees);
- ROM Pelvic Tilt (Range of Motion of Pelvic Tilt): calcu-

<table>
<thead>
<tr>
<th>Number of children</th>
<th>HSP group</th>
<th>SD group</th>
<th>Control group</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age (years)</td>
<td>10.07 (5.56)</td>
<td>8.59 (4.27)</td>
<td>10.50 (5.22)</td>
</tr>
<tr>
<td>Height (cm)</td>
<td>135.89 (12.99)</td>
<td>125.71 (16.90)</td>
<td>134.31 (7.11)</td>
</tr>
<tr>
<td>Weight (kg)</td>
<td>36.10 (13.96)</td>
<td>30.06 (14.66)</td>
<td>33.54 (10.39)</td>
</tr>
</tbody>
</table>

* = p-value < 0.05, comparison of HSP group and SD group; † = p-value < 0.05, compared with healthy subjects.
lated as the difference between the maximum and minimum values of the pelvic tilt angle, it represents the amplitude of movement of the pelvic joint in the sagittal plane (degrees).

**Hip flex-extension**
- **HIC** (Hip angle at Initial Contact): value of hip angle at initial contact (degrees);
- **HmSt** (Hip minimum angle in Stance): minimum hip angle value in the stance phase, which represents the peak of hip extension during the stance phase (degrees);
- **HMSw** (Hip Maximum angle in Swing): peak of hip angle in the swing phase, which represents the maximum hip flexion capacity during the swing phase (degrees).

**Knee flex-extension**
- **KIC** (Knee angle at Initial Contact): value of knee angle at initial contact (degrees);
- **KmSt** (Knee minimum angle at Midstance): minimum knee flexion value in midstance, which represents the maximum knee extension during the stance phase (degrees);
- **KMSw** (Knee Maximum angle in Swing): peak of knee flexion in the swing phase, which represents the maximum capacity of knee flexion during the swing phase (degrees).

**Ankle joint and foot**
- **AIC** (Ankle angle at Initial Contact): value of the ankle dorsi-plantarflexion angle at the moment of initial contact (degrees);
- **AMSt** (Ankle Maximum angle in Stance): peak of ankle dorsiflexion during stance phase, which represents the maximum ankle dorsiflexion capacity during the midstance phase (degrees);
- **ATO** (Ankle angle at Toe-Off): peak of ankle plantarflexion at the beginning of toe-off, which represents the maximum capacity of ankle plantarflexion at the end of stance phase (degrees);
- **AMSw** (Ankle Maximum angle in Swing): peak of ankle dorsiflexion during the swing phase, which represents the maximum capacity of ankle dorsiflexion during the swing phase (degrees);
- **Mean Foot Progression**: mean value of the foot progression angle, that is the angle between the foot vector and the line of progression, during the stance phase. This value represents the orientation (extra-intra rotation) of the foot with respect to the line of progression (degrees).

**Kinetics:**
- **Ankle power**
  - **min AP** (minimum value of Ankle Power): minimum value of absorbed ankle power in early stance and midstance, when muscle is contracting eccentrically and absorbing energy (minimum value of negative ankle power; W/kg);
  - **max AP** (maximum value of Ankle Power): maximum value of generated ankle power during terminal stance, when muscle is contracting and performing positive work. This value represents the peak of push-off ability at the end of the stance phase (maximum value of positive ankle power during terminal stance; W/kg).

The above indices were computed for each subject and mean values and standard deviations for the study groups and the control group were computed thereafter.

**Results**

Age, body weight, and height were not found to differ significantly among the HSP, SD and healthy subjects, as reported in Table I.

The ASH score ranged from 1 to 3.2 points in the HSP patients and from 1.7 to 2.8 points in the SD patients for both lower extremities; the GMFM ranged from 204 to 261 points in the HSP patients and from 163 to 256 points in the SD patients. No significant differences were found between the clinical features of the pathological groups.

Tables II, III and IV (over) report the mean values (and standard deviations) of all the GA parameters considered in this study, both kinematic and kinetic, in the HSP and SD groups and in the CG.

**Spatio-temporal parameters**

The analysis of spatio-temporal parameters (Table II) revealed no statistical differences between the two patho-
logical groups: when compared to the CG, the HSP and SD groups showed a % Stance that was close to normal, a lower mean Velocity (of progression), higher Step width values, and a reduced Anterior step length.

**Kinematic parameters**

As regards the kinematic parameters (Table III), no statistically significant differences were found between HSP and SD patients in relation to the pelvic joint. However, compared to the healthy subjects, both groups were characterised by a statistically significantly increased Mean Pelvic Tilt and by a significantly higher ROM Pelvic Tilt during walking in the sagittal plane. The hip joint was excessively flexed throughout the gait cycle in both groups with respect to normal range, but no statistically significant differences emerged between the HSP and SD groups in any of the parameters considered (HIC, HmSt and HMSw).

The knee flex-extension plot revealed that both the pathological groups presented excessive knee flexion at initial contact (KIC), with the HSP patients recording values significantly higher than those of the SD group. During midstance (KmSt) and in the swing phase (KMSw), both groups had knee angle values close to those of the CG.

In both the HSP and the SD groups, a significant percentage of patients displayed a tendency for knee hyperextension during midstance (50% in HSP and 56% in SD patients), and this was found to be significantly longer in the HSP subjects compared to the SD patients (HSP: 0.40±0.15 s; SD: 0.28±0.12 s; p<0.05).

Analysis of the ankle kinematics showed that the HSP group exhibited a pattern closer to normality than the SD patients, recording a normal ankle position at initial contact (AIC), and a better capacity of dorsiflexion in the stance (AMSst) and in the swing (AMSwf) phases. No significant differences were found in ankle joint position during terminal stance (ATO). The foot angle in the transverse plane (Mean Foot Progression) showed a normal foot orientation in the HSP group, while the SD patients, showing a statistically significant difference from the HSP and normative data, walked with their feet excessively extra-rotated.

### Table III - Mean (and standard deviation) values of gait analysis kinematic parameters in patients with hereditary spastic paraplegia (HSP), patients with spastic diplegia (SD) and healthy control subjects.

<table>
<thead>
<tr>
<th>Parameter</th>
<th>HSP group</th>
<th>SD group</th>
<th>Control group</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Pelvic tilt (in degrees)</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Mean Pelvic Tilt</td>
<td>18.19 (4.74)†</td>
<td>19.09 (6.63)†</td>
<td>8.85 (4.27)</td>
</tr>
<tr>
<td>ROM Pelvic Tilt</td>
<td>7.30 (3.53)†</td>
<td>8.76 (5.96)†</td>
<td>1.60 (3.64)</td>
</tr>
<tr>
<td><strong>Hip flex-extension (in degrees)</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Angle at Initial Contact</td>
<td>43.51 (8.74)†</td>
<td>38.28 (9.64)†</td>
<td>29.51 (4.40)</td>
</tr>
<tr>
<td>Minimum angle in Stance</td>
<td>-1.21 (8.26)†</td>
<td>-0.83 (8.70)†</td>
<td>-8.70 (6.41)</td>
</tr>
<tr>
<td>Maximum angle in Swing</td>
<td>48.24 (9.68)†</td>
<td>43.94 (8.67)†</td>
<td>30.71 (5.23)</td>
</tr>
<tr>
<td><strong>Knee flex-extension (in degrees)</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Angle at Initial Contact</td>
<td>27.24 (6.17)†</td>
<td>19.04 (5.11)†</td>
<td>6.72 (5.53)</td>
</tr>
<tr>
<td>Minimum angle at Midstance</td>
<td>0.48 (10.63)</td>
<td>-0.94 (11.17)</td>
<td>4.35 (5.26)</td>
</tr>
<tr>
<td>Maximum angle in Swing</td>
<td>56.54 (11.84)</td>
<td>55.83 (9.79)</td>
<td>60.54 (4.74)</td>
</tr>
<tr>
<td><strong>Ankle joint and foot (in degrees)</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Angle at Initial Contact</td>
<td>0.95 (5.40)*</td>
<td>-6.49 (5.78)†</td>
<td>0.40 (3.81)</td>
</tr>
<tr>
<td>Maximum angle in Stance</td>
<td>13.66 (6.17)*</td>
<td>6.02 (7.45)†</td>
<td>12.27 (5.53)</td>
</tr>
<tr>
<td>Angle at Toe-Off</td>
<td>-12.38 (14.34)</td>
<td>-11.91 (9.82)</td>
<td>-11.06 (5.23)</td>
</tr>
<tr>
<td>Maximum angle in Swing</td>
<td>7.49 (6.59)*</td>
<td>-0.16 (7.21)†</td>
<td>5.78 (6.46)</td>
</tr>
<tr>
<td>Mean Foot Progression</td>
<td>-10.73 (6.42)*</td>
<td>-18.41 (6.53)†</td>
<td>-12.72 (4.26)</td>
</tr>
</tbody>
</table>

* = p-value < 0.05, comparison of HSP group and SD group; † = p-value < 0.05, compared with healthy subjects.

### Table IV - Mean (and standard deviation) values of gait analysis kinetic parameters in patients with hereditary spastic paraplegia (HSP), patients with spastic diplegia (SD) and healthy control subjects.

<table>
<thead>
<tr>
<th>Parameter</th>
<th>HSP group</th>
<th>SD group</th>
<th>Control group</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ankle Power (W/kg)</td>
<td>-0.72 (0.20)</td>
<td>-0.59 (0.44)</td>
<td>-0.50 (0.29)</td>
</tr>
<tr>
<td>Minimum of absorbed power</td>
<td>-0.91 (0.48)†</td>
<td>1.21 (1.07)†</td>
<td>3.75 (1.89)</td>
</tr>
</tbody>
</table>

* = p-value < 0.05, comparison of HSP group and SD group; † = p-value < 0.05, compared with healthy subjects.
Kinetic parameters

No statistical differences in ankle power parameters (Table IV) emerged between the HSP and SD groups: the patients of both groups demonstrated minimum absorbed power values (min AP) in the early stance and midstance phases that were close to those of the CG and a push-off ability at the end of the stance phase (Max AP) that was statistically significantly reduced compared to normative data.

Discussion

The aim of this study was to evaluate, by means of GA, gait strategies in patients affected by HSP and in those with SD secondary to CP, focusing on the differences between the two groups of patients during walking. Subjects affected by HSP, in fact, resemble patients who have a mild form of SD, but since HSP is a progressive neurodegenerative disorder (as opposed to a stable condition, like SD), and also in view of the different kinematic patterns highlighted in this study, specific therapeutic programmes are needed for the two conditions. So the main clinical and rehabilitative concern is to find an accurate way of distinguishing between these two pathologies; GA was applied to these patients with precisely this aim, in order to obtain information in addition to that which can be obtained from classical clinical evaluations and that may be useful for identifying the correct treatment for the patient. The results of this study showed that the two groups share certain characteristics of gait, particularly with regard to spatio-temporal parameters and proximal joint indices. They revealed, in fact, spatio-temporal parameters (Velocity of progression, Step width and Anterior step length) that differed from normal values: these parameters delineated a cautious abnormal gait in both groups of patients, which probably reflects their attempt to achieve equilibrium and stability. No significant differences were found in pelvis and hip joint position in the sagittal plane: indeed, all the patients exhibited, compared to the healthy group, anterior pelvic tilt with high joint excursion during walking and excessive hip flexion during the whole gait cycle. The analysis of knee kinematics provided the most significant information from the perspective of characterising gait pattern in HSP and establishing a clear differentiation between HSP and SD patients: the subjects with HSP showed greater knee flexion at initial contact and, where present, significantly longer-lasting knee hyperextension in midstance compared to the SD group. As stance duration is similar in the two pathological groups, the prolonged duration of the knee hyperextension in the HSP patients represents a distinctive aspect of the motor strategy of these subjects compared to the SD patients.

Parameters extracted from ankle dorsi-plantarflexion and foot progression plots showed that the HSP group exhibited mean values that were close to normality, whereas the SD patients generally showed excessive plantarflexion and external foot rotation throughout the gait cycle.

No statistical differences were found in the ankle kinetic parameters: both groups were in fact characterised by a reduced push-off ability at the end of the stance phase, probably due to spasticity of the calf muscle. The results obtained highlighted that ankle and especially knee kinematic strategies are aspects on the basis of which it is possible to distinguish between the gait patterns of HSP and SD patients. In particular, with regard to the knee joint, it is important to draw attention to the different biomechanical features leading to knee hyperextension during the midstance phase. In SD patients, knee hyperextension is linked to an increase of the plantarflexion/knee extension couple: the ground reaction force falls in front of the knee and generates an extensor moment. Consequently the hip- and knee-extensor muscles must remain active to prevent these joints from collapsing into flexion (11). In HSP patients, on the other hand, knee hyperextension, which lasts longer and is not connected to ankle plantarflexion, could be due to a compensatory knee stabilisation strategy; indeed, in the absence of this biomechanical condition, the muscles directly connected to the knee joint, which are hyposthenic, would not be able to work eccentrically in order to avoid the knee from giving way during walking. This could be the reason why patients affected by HSP, treated with intrathecal baclofen, need a much lower drug dosage than SD patients. Moreover, if in SD patients the use of an ankle foot orthosis is able to reduce the knee hyperextension by diminishing the plantarflexion/knee-extension couple, in HSP patients a reduction of the knee’s ability to hyperextend due to use of an orthosis could interfere with the patient’s knee stabilisation strategy. In the light of these two different biomechanical conditions during gait, in particular at the level of the proximal joints, it is clear that the two pathologies demand different therapeutic and rehabilitative interventions. For this reason, it is very important, from a clinical perspective, to be able to distinguish clearly and accurately between them. The results of this study demonstrated that GA furnishes clinicians with information in addition to that which can be obtained from traditional evaluations, allowing them to identify the most appropriate course of treatment, neurosurgical or pharmacological. Reduction of the spasticity in SD can lead, in fact, to a significant gait improvement, but the same treatment in HSP may weaken the muscles of the lower limbs interfering with compensation strategies, such as knee hyperextension.

References