Original article

3D gait analysis in patients with hereditary spastic paraparesis and spastic diplegia: A kinematic, kinetic and EMG comparison

Luigi Piccinini a, Veronica Cimolin b,*, Maria Grazia D’Angelo a, Anna Carla Turconi a, Marcello Crivellini b, Manuela Galli b,c

a IRCCS “E. Medea”, “La Nostra Famiglia” Association, Bosisio Parini, Lecco, Italy
b Bioeng. Dept. Politecnico di Milano, P.zza Leonardo da Vinci 32, 20133 Milano, Italy
c IRCCS “San Raffaele Pisana” San Raffaele SPA, Roma, Italy

ABSTRACT

The predominant clinical feature of patients with Hereditary Spastic Paraparesis (HSP) is gait disturbance owing to spasticity and weakness of the lower limbs; the spasticity in early-onset disease (infancy or childhood) often cannot be distinguished from mild form of spastic diplegia (SD). The aim of this study was to quantify the gait strategy in HSP and SD children, focusing on the differences between groups as concerns functional limitation during gait.

9 HSP and 16 SD children were evaluated using Gait Analysis; kinematic and kinetic parameters and EMG pattern during walking were identified and calculated to compare the two gait strategies. The results revealed that these two pathologies are characterised by different gait strategies. In particular we found that knee joint, in terms of kinematics and kinetics, and rectus femoris pattern represent discriminatory aspects in order to compare and differentiate gait patterns of HSP and SD children.

The findings strongly support the issue that HSP and SD patients need individualised therapeutical program, either neurosurgical or pharmacological treatment, based on the quantification of gait deficiencies and in order to address the peculiarity of their motor limitations and to prevent the onset of compensatory strategies.

© 2010 European Paediatric Neurology Society. Published by Elsevier Ltd. All rights reserved.

1. Introduction

Hereditary Spastic Paraparesis (HSP) is a heterogeneous group of neurodegenerative disorders in which the predominant clinical feature is gait disturbance owing to spasticity and weakness of the lower limbs. The spasticity in early-onset disease (infancy or childhood) often does not progress significantly and cannot be distinguished from mild form of spastic diplegia secondary to Cerebral Palsy (CP), except when a family history is elicited.1,2

Up to 25% of affected patients are asymptomatic, and so it is possible a patient has only subclinical manifestations,
therefore escaping diagnosis. The gait abnormalities observed in children with HSP are the earliest clinical finding because classic features for spastic paraplegia are not sometimes seen in these patients; for this reason in these patients a careful and complete clinical evaluation, including gait evaluation, is crucial.

Controversially, there is a lack of detailed quantitative analyses of gait in these patients. Most of the evaluations present in literature were conducted using clinical-functional scales and kinematics of main lower limb joints; in addition they were focused mainly on adult patients. As for children with HSP, only one quantitative study investigating the biomechanical strategy during gait is available to our knowledge.

Cimolin et al. quantified the biomechanical strategy in HSP children during gait, comparing their pattern to children who have a mild form of spastic diplegia secondary to CP, as paediatric patients with HSP often resemble diplegic children. Children with HSP, were compared to children with spastic diplegia using Gait Analysis (GA). The main results of this study were the following:

a) HSP and SD were found similar in spatio-temporal and kinematic parameters at proximal joints.

b) The most significant differences were at knee and ankle joints.

c) They both showed tendency to knee hyperextension in midstance, but hyperextension timing was longer in HSP.

In this study, however, the evaluation of gait strategy was focused only on spatio-temporal parameters and kinematic and scanty information (only about ankle power) were given in respect of kinetics and EMG pattern during gait. In addition, a heterogeneous group of spastic paraparesis patients were selected, without considering the genetic origin. From a clinical perspective it is very important to evaluate the gait pattern in these patients using both clinical scores and instrumental quantitative measures, not only with regard to kinematics but also kinetic and EMG data of mainly lower limb joints. The assessment of gait pattern using only spatio-temporal parameters and kinematics is not enough because they give a limited evaluation of patient’s walking ability; for this reason the integration of these data with kinetics and EMG is useful for better investigating the joint reactions, moments, powers and muscular activity. In this way it is possible to assess the mechanisms that either control or produce movement, thus potentially developing a more comprehensive understanding of motion and providing insight not only into the ‘how’ (kinematics), but also into the ‘why’ (kinetics) of the movement we observe. In addition, dynamic EMG provides the timing and action of muscles that are the prime movers of body segments and bones. Understanding the activity of the muscle as well as the other forces acting on a moving body is critical to understand the root causes of a gait abnormality.

In this context, starting from previous results, the aims of this study are: (a) to quantify the functional limitation of children with genetically defined HSP, using 3D Gait Analysis (GA), in terms of spatio-temporal parameters, kinematics, kinetics and EMG; (b) to identify and calculate the differences between genetically defined HSP and spastic diplegia (SD) secondary to CP, using quantitative parameters obtained from GA data (kinematic, kinetic and EMG data).

2. Materials and methods

2.1. Subjects

9 patients with the clinical diagnosis of Hereditary Spastic Paraplegia (HSP; age: 8.9 ± 3.1 years; height: 129.9 ± 9.9 cm; weight: 36.1 ± 8.9 Kg) and 16 patients with Spastic Diplegia (SD; age: 11.9 ± 2.4 years; height: 125.7 ± 7.3 cm; weight: 30.1 ± 7.6 Kg), secondary to Cerebral Palsy, were evaluated in this study.

We studied a referral cohort of HSP patients, according to the following selection criteria: a clinical diagnosis of spastic paraplegia in absence of structural/spinal cord/cerebral disorders, demyelinating, metabolic or inflammatory disorders (particularly for the sporadic forms) and, for the familial forms presence of a positive family history of “gait disturbances” and/or gene mutations (most commonly in the SPG4 and SPG7 genes). All patients underwent metabolic screening; they all had a brain and a spinal cord MRI, reported as normal. All patients have been genetically defined: 4 with mutation in the mitofusin gene, 3 with a mutation in the atlastin gene (inherited by the father) and 2 with a mutation in the spastin gene (a “de novo” mutation, being the parents negative for any mutation in the spastin gene).

Selection criteria for patients with SD were a physician diagnosis of spastic diplegia of Type III according to Rodda’s classification, with a mild spasticity of lower limbs joints, no history of cardiovascular disease and no previous surgery or other significant treatments for spasticity.

All patients were able to walk independently without the use of crutches, walkers or braces.

A control group of 15 non-affected subjects (CG; age: 9.2 ± 5.7 years; height: 130.3 ± 7.1 cm; weight: 33.5 ± 9.4 Kg) was included. Selection criteria for this second group included no prior history of cardiovascular, neurological or musculo-skeletal disorders. They exhibited normal ROM and muscle strength, and had no apparent postural and motor deficits.

All subjects were volunteers and their parents gave their written consent to the children’s participation in this research, in accordance with the local ethical committee requirements.

2.2. Data collection

The complete evaluation consisted of three components: clinical examination, video-recording and 3D Gait Analysis (GA).

The “Ashworth scale of muscle spasticity” (ASH) and the Gross Motor Function Measure (GMFM) were assessed in the clinical examination.

The ASH scale evaluates the severity of spasticity calculated as the mean value of spasticity in both lower limbs. The GMFM measures the child’s overall functional abilities and it consists of 88 items, divided into the following sections: 1) lying and rolling; 2) sitting; 3) crawling and kneeling; 4)
standing; 5) walking, running and jumping. Each section contributes to the total GMF score (range: 0–264). The GMFM demonstrated excellent psychometric properties in children with CP but in this last decade its validity was demonstrated also in other pathologies, such as Osteogenesis Imperfecta, Down Syndrome and Traumatic Brain Injury, supporting the use of the GMFM as an evaluative measure of gross motor function in patients with motor disabilities. According to literature and our experience we considered a valuable assessing method also in HSP patients.

Muscuary strength was evaluated according to Medical Research Council (MRC) Scale for Muscle Strength of knee extensor (quadriceps). This scale ranges from grade 0 (no movement observed) to grade 5 (muscle contracts normally against full resistance).

GA was conducted using an optoelectronic system with passive markers (ELITE2002, BTS, Milan, Italy) working at a sampling rate of 100 Hz, for kinematic movement evaluation, a 6-channels surface EMG system (TeleEMG, BTS, Milan, Italy) for muscles electromyographic signals monitoring, and a Video system synchronic with the optoelectronic and EMG systems (BTS, Milan, Italy).

After collecting some anthropometric measures (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 and thickness of the pelvis), passive markers were placed at special points of reference, directly on the subject’s skin, as described by Davis. For the surface EMG recording, bipolar Ag/AgCl surface electrodes pairs with a diameter of 10 mm and an inter-electrode spacing of 22 mm were placed bilaterally on clean, shaven skin overlying the rectus femoris (RF). The SENIAM recommendations for surface EMG were followed for electrode placement. The ground electrode was placed overlying the tibial tuberosity. EMG signals were pre-amplified, band-pass filtered (10–700 Hz) at a sampling rate of 2520 Hz, but not processed further. After subject’s preparation, patients walked barefoot at their self-selected speed with a walking speed of 25–20 km/h.

Some parameters were identified and calculated from kinematic and kinetic data: spatio-temporal parameters, angles joint values (pelvis, hip, knee and ankle in the frontal/sagittal/ transversal planes) in specific gait cycle instant and peak values of joint moments and powers (hip, knee and ankle) angles.

The kinematic and kinetic data are presented as average values for each group and were compared to values of the control group.

As concerns EMG data, abnormalities of EMG activity and timing were required for the definition of abnormal muscle activity. The criteria were derived from our database on normal subjects and from literature and were as follows:

1) Activity and timing:
   a) Knee extensor activity in mid or late stance;
   b) Continuous knee extensor activity during swing phase.

2.4. Statistics

A one-way between groups analysis of variance (ANOVA) was applied for statistical analysis, the assumptions of the ANOVA model were tested by evaluating the fit of the observed data to the normal distribution (Kolmogorov–Smirnov test) and the homogeneity of variances (Levene’s test). Specific effects were evaluated by means of the post-hoc comparisons of means (Bonferroni test). Null hypotheses were rejected when probabilities were below 0.05.

3. Results

Age, body weight, and height were not significantly different among pathological and healthy participants.

All the patients were able to complete both clinical and instrumental evaluation with GA.

The demographic data and clinical measures of the participants to this study were

3.1. Clinical examination

The ASH ranged from 1.1 to 3.2 points in HSP patients and from 1.9 to 3.1 points in SD patients for both lower extremities; the GMFM ranged from 204 to 260 points in HSP patients and from 170 to 250 points in SD patients.

The MRC Scale for Muscle Strength of quadriceps revealed that HSP patients were characterised by grade 1 and 2, while SD patients grade 3 and 4. No significant differences were found between pathological groups as concern clinical features.

3.1.1. 3D Gait Analysis

In Tables 1–3, the mean values (±standard deviation) of all GA parameters considered in this study for the HSP, DS groups and for Control Group were reported.

Please cite this article in press as: Piccinini L, et al., 3D gait analysis in patients with hereditary spastic paraparesis and spastic diplegia: A kinematic, kinetic and EMG comparison, European Journal of Paediatric Neurology (2010), doi:10.1016/j.ejpn.2010.07.009
Table 1 – Mean (standard deviation) values of spatio-temporal parameters for HSP group (patients with Hereditary Spastic Paraparesis), SD group (patients with Spastic Diplegia) and for healthy subjects (Control Group).

<table>
<thead>
<tr>
<th>Spatio-temporal parameters</th>
<th>HSP Group</th>
<th>SD Group</th>
<th>Control Group</th>
</tr>
</thead>
<tbody>
<tr>
<td>% Stance (% gait cycle)</td>
<td>60.3 (2.6)</td>
<td>59.6 (2.7)</td>
<td>59.0 (1.9)</td>
</tr>
<tr>
<td>Velocity (m/s)</td>
<td>0.9 (0.2)a</td>
<td>1.1 (0.1)a</td>
<td>1.3 (0.2)</td>
</tr>
<tr>
<td>Step Width (mm)</td>
<td>126.5 (32.4)b</td>
<td>141.1 (41.2)a</td>
<td>97.4 (22.3)</td>
</tr>
<tr>
<td>Anterior step length</td>
<td>0.4 (0.3)a</td>
<td>0.4 (0.2)a</td>
<td>0.8 (0.2)</td>
</tr>
</tbody>
</table>

a p-value < 0.05, compared with healthy subjects.

3.2. Spatio-temporal parameters (Table 1)

Both pathological groups were characterised by normal duration of stance phase, shorter anterior step length, lower velocity of progression and larger step width when compared to control group. No statistical differences were found among HSP and SD groups.

3.3. Kinematic parameters (Table 2)

With regard to pelvis, no significant differences were found in the three planes of movement in terms of range of motion (ROM Pelvic Tilt, ROM Pelvic Obliquity and ROM Pelvic Rotation indices). Both groups were characterised by higher Pelvic Tilt excursion and abnormal ROM of Pelvis on the frontal and transversal planes if compared to control group.

The hip joint exhibited excessive flexed position during the whole gait cycle (HC and HmSt indices) in HSP and SD patients, with similar values.

As concerns hip intra-extrarotation, the SD subjects were characterised by hip intra-rotation throughout the entire gait cycle higher than HSP and CG. HSP groups revealed hip position on the transversal plane close to normality.

The knee flex-extension plot revealed that both pathological groups presented excessive knee flexion at initial contact (KIC index), mean values quite normal during midstance (KmSt index) and lower values of knee flexion in swing phase (KMSw index) than CG (Fig. 1).

Even if the mean values of knee angle in stance are similar to healthy individuals, it is important to highlight that in pathological groups, both in HSP and in SD group, a significant percentage of patients revealed the tendency to knee hyperextension during midstance (70% in HSP and 65% in SD patients). A significant difference was found in term of duration of knee hyperextension: HSP subjects were characterised, in fact, by significant longer phase of knee hyperextension during midstance respect to SD patients (HSP: 0.32 ± 0.07 s; SD: 0.18 ± 0.08 s; p < 0.05).

Analysis of ankle kinematics showed that in HSP group patients generally exhibited quite normal position of ankle during the whole gait cycle. On the contrary, the SD patients displayed excessive plantarflexion at initial contact (AIC index) and reduced ability in dorsiflexion during stance (AMSt index) and swing phase (AMSw index).

Please cite this article in press as: Piccinini L, et al., 3D gait analysis in patients with hereditary spastic paraparesis and spastic diplegia: A kinematic, kinetic and EMG comparison, European Journal of Paediatric Neurology (2010), doi:10.1016/j.ejpn.2010.07.009
Knee kinetics exhibited significant differences in terms of moment and power. As for knee moment, HSP patient generally presented higher flexor moment in midstance than SD and healthy subjects. Knee power revealed that HSP patients were characterised by a maximum value (generated power) lower than SD, that was close to CG; in terms of minimum power, both groups displayed higher values of this parameter, but HSP patients were characterised by more abnormal values than SD (Fig. 1).

Foot angle in transversal plane (Mean Foot Progression index) pointed out quite normal foot orientation in both pathological groups.

3.4. Kinetic parameters (Table 3)

No statistical differences were found in terms of hip moment (max extension moment) and power at initial stance; both groups revealed higher values than CG.

![Fig. 1](image1.png)

Fig. 1 – Knee flex-extension angle, knee moment and knee power of a trial of two patients (solid line: HSP patient; dashed-line: SD patient) and normative range of control group (thick lines) are reported.

![Fig. 2](image2.png)

Fig. 2 – Hip flex-extension, knee flex-extension and ankle dorsi-plantarflexion plot of a trial of two patients (solid line: HSP patient; dashed-line: SD patient) and normative range of control group (thick lines) are reported.
As concerns ankle joint, both in terms of moment and power, no statistically different patterns were displayed by HSP and SD patients: they both revealed lower values of peak in plantarflexor moment during terminal stance, quite normal values of minimum absorbed power in early stance and midstance and more limited values of maximum ankle power generation at push-off, if compared to normative data.

In Fig. 2 an example of hip flex-extension, knee flexion and ankle dorsi-plantarflexion plots were reported for a patient with HSP and one with SD in order to better detect the differences between the two pathological states.

3.5. EMG data

In terms of EMG activity of rectus femoris, two different situations were found in the pathological groups: while most of HSP patients (14/18 side: 78% of sides) generally showed a very low activation of rectus femoris during all gait cycle, displaying the presence of muscular hypotension, most of SD subjects (26/32 sides: 81% of sides) was characterised by higher knee extensor activation in stance phase than HSP patients, evidencing the presence of rectus femoris spasticity (Fig. 3) frequently coactive with hamstrings in patients with CP.

4. Discussion

The aim of this study was the quantification of functional limitations in terms of gait pattern in patients affected by HSP and SD, focusing on the differences between groups, using GA (spatio-temporal parameters, kinematics, kinetics and EMG).

Children with HSP often resemble children who have a mild form of spastic diplegia secondary to CP, but they differ as concern motor limitations. While gait pattern in SD secondary to CP has been sufficiently addressed in order to provide evidence-based rehabilitation strategies, there is scanty evidence in the quantification of gait patterns in HSP subjects and subsequently rehabilitation programs fail to have been implemented on the basis of objective functional data in HSP. Conversely, we focused our attention on gait, and a quantitative description of the characteristic gait manifestations of these pathologies has been developed, conducting an in-depth investigation into the diversities in their gait patterns in terms of kinematics, kinetics of main lower limb joints and EMG.

Firstly, our data showed that no significant differences were found in terms of clinical features, even if the HSP seemed to be a slightly more compromised than SD, especially in terms of MRC of quadriceps (HSP were characterised by grade 1 and 2, while SD grade 3 and 4); this means that from a clinical point of view the two pathological states were similar and for this reason the biomechanical quantification was necessary.

As concerns spatio-temporal parameters and kinematics, the results of this study are in line with literature. Both groups were in fact characterised by some common peculiarities during walking, in particular in terms of spatio-temporal parameters and indexes on proximal joints. They revealed, in fact, spatio-temporal parameters different from normality, as concern velocity of progression, step width and anterior step length. These parameters indicate a cautious, abnormal gait in both groups, aimed to provide stability in subjects characterised by balance-related gait features.

No significant differences were found at pelvis and hip joint position on sagittal plane: all patients exhibited pelvic tilt with high excursion during walking and excessive hip flexion during the whole gait cycle if compared to healthy group. Conversely to previous study, we integrated the analysis considering the gait pattern in the frontal and coronal planes, too: while no differences were found in terms of pelvis, hip angle in the transversal plane demonstrated that HSP revealed a more physiological position than SD patients, which walked with more intra-rotated hip joint.

This different hip position may be due to a physiological correction of femur anteversion in the first years of life in HSP patients compared to a permanence of neonatal femur anteversion in SD patients where neurological anomalies are present at birth. In this study we considered only joint clinical evaluation for femoral anteversion; according to literature we considered that the clinical method for the determination of the degrees of femoral anteversion is superior to radiographic techniques in children who have not had a previous operation about the hip, as our patients.

As already demonstrated, knee kinematics provides the most significant information in order to characterise gait pattern in HSP and to make a clear differentiation between HSP and SD patients: subjects with HSP revealed knee more flexed at initial contact and, where present, knee hyperextension in midstance significantly longer than SD. As stance duration is similar in the two pathological groups, the prolonged timing of knee hyperextension in HSP patients represents a considerable feature of motor strategy of these subjects in comparison with SD patients.

Parameters extracted by ankle dorsi-plantarflexion and foot progression plots showed that HSP group exhibited an ankle strategy close to normality, while SD were characterised by an increased plantar flexion and reduced dorsiflexion throughout the gait cycle.
No statistical differences were found as concern hip kinetic parameters: both groups were characterised by prolonged and greater than normal hip-extensor moment and hip power generation in stance. This kinetic pattern is directly connected to increased hip flexion throughout the gait cycle that we observed in HSP and SD patients.\textsuperscript{18}

On the contrary, significant differences were found in terms of knee kinetics between two analysed pathological groups. We observed that HSP patients were characterised by excessive knee flexor moment in midstance, limited value of maximum generated power if compared to SD, that was close to CG; in terms of minimum power, representative of power absorption, both groups displayed higher values of this parameter, but HSP patients were characterised by slightly increased power absorption than SD. These results are directly connected to the presence of knee hyperextension in stance; while in SD patients this pattern is due to plantarflexor spasticity contributing to an excessive plantarflexion/knee extension couple in HSP it may be related to rectus femoris weakness/hypoactivation, as demonstrated by EMG pattern.

No statistical differences were found in the ankle kinetics: both HSP and SD patients were characterised by a reduced push-off ability in terminal stance, maybe connected to a generalised spasticity of the calf muscle, condition frequently present in these patients, both in HSP\textsuperscript{19} and SD.\textsuperscript{18}

The obtained results highlighted that not only knee kinematics, as previously demonstrated, but also knee kinetics and rectus femoris pattern represent discriminatory aspects in order to compare and differentiate gait patterns of HSP and SD patients. In particular, it is important to point out the different biomechanical features which lead to knee hyperextension during midstance in the two pathological groups. In SD children knee hyperextension is connected to increase of plantarflexion/knee extension couple: the ground reaction force falls in front of the knee and generates a knee external extensor moment in stance. Consequently kinetics shows a peak of internal eccentric flexor moment.\textsuperscript{18} In HSP children knee hyperextension, longer and not connected to ankle plantarflexion, represents a compensatory strategy for knee stabilization. The patients with HSP showed a quadriiceps avoidance pattern with a reduced knee extensor weakness on MRC, as demonstrated by our data, slightly decreased extensor moment during loading response if compared to healthy individuals, even if not statistically significant, and the absence of quadriiceps activity, as displayed by the EMG signal.

If this biomechanical condition does not happen, muscles directly connected to knee joint, as rectus femoris which is characterised by hypostenia, would not able to work eccentrically in order to avoid the knee collapse during walking.

These results may be useful from a clinical viewpoint as 3D gait analysis can deeply investigate the gait strategy in patient with genetically defined HSP.

\section*{References}