Gait & Posture 33 (2011) 556–561

Contents lists available at ScienceDirect

Gait & Posture

journal homepage: www.elsevier.com/locate/gaitpost

Gait analysis may help to distinguish hereditary spastic paraplegia from cerebral palsy

Sebastian I. Wolf [a,*], Frank Braatz [a], Dimitrios Metaxiotis [a,b], Petra Armbrust [a], Thomas Dreher [a], Leonhard Döderlein [a,c], Ralf Mikut [d]

[a] Department of Orthopaedic Surgery, University of Heidelberg, Schlierbacher Landstr. 200a, 69118 Heidelberg, Germany
[b] Orthopaedic Department, Papageorgiou Hospital, Thessaloniki, Greece
[c] Behandlungszentrum Aschau, Germany
[d] Institute for Applied Computer Science, Karlsruhe Institute of Technology (KIT), Germany

ARTICLE INFO

Article history:
Received 27 July 2010
Received in revised form 13 January 2011
Accepted 15 January 2011

Keywords:
Hereditary spastic paraplegia
Gait analysis
Automated methods
Classification
Feature extraction
Pattern recognition

ABSTRACT

Hereditary spastic paraplegia (HSP) designates a group of genetic disorders typically leading to spasticity in the lower limbs and consequently to gait disorders. Although the symptoms are similar to those of cerebral palsy (CP), the correct diagnosis is important for treatment recommendations as one condition is progressive in nature whereas the other is not. Due to the heterogeneity of HSP, genetic testing is complex and in some genetic forms still not possible. The aim of this study was, therefore, to investigate if instrumented 3D-gait analysis could help distinguish between these two conditions.

The gait pattern of 29 patients with HSP was compared with that of 29 patients with CP who were matched in age, sex, and the extent of gait disturbance and also to 29 typically developing subjects for reference. More than 3000 gait parameters were evaluated for their relevance to classify patients into diagnostic groups. Cluster analysis revealed that these gait features may classify only subgroups of symptoms as the gait pattern is very heterogeneous within each diagnosis group. However, prolonged hip extension, knee extension, and ankle plantar flexion were identified as indicators for HSP. In addition, large trunk tilt velocities appear unique in some cases of HSP. These indicators in gait pattern may contribute in establishing the diagnosis of HSP, which is important in predicting outcome when planning surgical treatment for functional improvements in these patients.

1. Introduction

Hereditary spastic paraplegia (HSP) designates a heterogeneous group of genetic disorders of which not all can yet be diagnosed by routine genetic testing [1]. In its "pure" form, HSP leads to progressive spasticity in the lower limbs due to pyramidal tract degeneration [2]. HSP is classified as complex when complicated by other neurological signs such as ataxia, mental retardation, dementia, extrapyramidal signs, visual dysfunction, or epilepsy [3]. Pure HSP represents around 70% of the cases and is typically inherited in an autosomal dominant manner [3,4]. In most cases, the phenotype includes slowly progressive spasticity in the lower limbs, resulting in gait deficiencies and loss of mobility starting in the second decade after the onset of symptoms. Clinically, the onset of this type of HSP can be from childhood through to late adult life, whereas more than half of mutation carriers do not develop symptoms until after the age of 30 years [5]. Although the clinical findings may vary, two groups of clinical types have been described based on the age at presentation: type I before and type II after 35 years of age [6]. With an onset in the first few years of life, combined with delayed motor milestones, the diagnosis is more suggestive of cerebral palsy (CP), particularly if the clinical picture is relatively static [5]. However, due to the progressive nature of the clinical symptoms in HSP, these patients are typically treated differently to patients with CP. Since genetic testing is not always possible and both the clinical status and the patients' history may not always be sufficient to establish the diagnosis, a detailed analysis of the gait pattern may help in the decision for an appropriate treatment.

Unlike CP, where gait has been studied in detail using 3D motion capture, only few studies have objectively described gait function in HSP. Braschinsky et al. [7] reported on reduced walking speed in 46 patients set in the context of reduced active hip flexion and abduction ROM as well as reduced ankle dorsiflexion ROM, which were determined clinically. Earlier, the group of Klebe found a reduced ROM in knee flexion and reduced walking speed due to reduced stride length and cadence along with an increase in step width in 22 adult patients (age 35–61 years) when compared to

* Corresponding author. Tel.: +49 6221 966724.
E-mail address: sebastian.wolf@med.uni-heidelberg.de (S.I. Wolf).

0966-6362/$ – see front matter © 2011 Elsevier B.V. All rights reserved.
doi:10.1016/j.gaitpost.2011.01.009

healthy controls [8]. The most detailed findings so far were reported in 15 children with HSP (aged 6–15 years) by comparing their gait to that of 40 patients with CP and 20 healthy control subjects with the aim to quantitatively assess differences in gait between the two conditions [9]. Temporospatial and kinematic parameters were found to be similar in the two patient groups but knee flexion at initial contact was particularly increased when compared to the CP group. Both groups tended to hyperextend the knee in mid-stance, however, a longer duration of hyperextension was observed in patients with HSP.

These first objective findings on gait characteristics in HSP and the clinical observation that some patients show a gait pattern atypical for CP suggest that objective gait observation may help in distinguishing the two conditions. Therefore, the aim of this retrospective study was to identify pathology-related gait patterns or characteristic features irrespective of the patient history. For this, a systematic approach based on both gait feature computation as described in [10] and on the use cluster analysis was employed. Instrumented gait analysis could then be used both for a better diagnostic work-up and for determining the best treatment options for the patient.

2. Patients

Thirty-five patients with clinical symptoms of HSP were seen between 1996 and 2008 at the outpatient clinics of our hospital. Instrumented 3D-clinical gait analysis (CGA) based on a conventional gait model [11] was conducted to establish further treatment recommendations. Of these patients, 29 (aged 5–63 years, 8f/21m) were included in the study since (a) gait data were reasonably reliable, i.e., data from five or more trials were measured and averaged, (b) video documentation was available, and (c) the diagnosis of HSP was confirmed by either a documented family history or by late age of onset (for HSP type II cases). The gross motor function score (GMFCS, [12]) in eight cases was classified as GMFCS 1, in another 18 patients as GMFCS 2, and in three patients as GMFCS 3. On their first visit, 24 subjects had not previously had any operations whereas five had received soft-tissue operations (calf muscles), among them two who also received proximal procedures (adductors, hamstrings, rectus femoris). More details about the subjects are given in Table 1.

Further, a group of 385 patients diagnosed with diplegic type CP who presented for their first visit in our hospital and who had also undergone CGA were scanned for the same inclusion criteria as in the HSP group. Sorted by age and sex, best matches to the HSP group were manually picked in a blinded manner only by the information on age, sex, and Gillette Gait Index (GGI) [13]. In this group seven cases were classified as GMFCS 1, in another 16 patients as GMFCS 2, and in six patients as GMFCS 3. It turned out (by accident) that, similarly to the HSP group, five of these 29 patients had also undergone soft-tissue operations (calf muscles), among them three who had also had proximal procedures, including one case of bony correction.

Hence the two groups showed very similar clinical backgrounds but with different diagnoses. For reference and testing reasons, a third group of typically developing subjects (NORM) was selected retrospectively out of the gait data base, aiming for best matches in age and sex.

3. Methods

For the instrumented gait analyses a Vicon 370 motion capture system was used during the years 1996–2001 and was then replaced by a Vicon 612 system applying a conventional gait model [11]. For each patient, the data of at least five strides of different trials were averaged. To assess gait patterns, a previously developed methodological modular framework was applied [10]. This framework formalizes the processing steps of data selection, gait parameter calculation, and evaluation, as well as classification according to the clinical problem. For these steps, several mathematical methods were selected and the validity of the approach was tested by applying it to the clinical problem of Botulinum Toxin-A treatment for spastic equinus [10]. The methodology is only briefly described here. An extended summary can be found in the electronic appendix. "Original time series", i.e., 3D

Table 1
Subject characteristics.

Triple	HSP group						CP group					Reference	
	Sex	Age	GGI	Walking aid	Previous surgeries	HSP type	Sex	Age	GGI	Walking aid	Previous surgeries	Sex	Age
1	m	5.6	72	No	No	I	m	4.8	89	No	No	m	5.0
2	f	6.2	132	No	No	I	f	6.0	111	No	No	f	5.9
3	m	8.2	131	No	No	I	m	7.7	98	No	No	m	7.9
4	m	8.7	360	No	No	I	m	8.7	303	No	No	m	8.7
5	m	9.0	305	No	No	I	m	9.3	304	No	No	m	9.1
6	f	9.1	277	No	No	I	f	8.8	266	No	No	f	9.4
7	m	9.5	1511	No	No	I	m	9.6	1584	No	No	m	9.1
8	m	9.9	45	No	No	I	m	10.3	137	No	No	m	10.2
9	f	11.9	606	No	No	I	f	11.7	581	No	No	f	11.4
10	f	12.1	271	No	Yes	I	f	11.7	279	No	No	f	11.7
11	m	14.0	100	No	No	I	m	13.3	128	No	No	m	11.7
12	m	14.2	118	No	No	I	m	14.4	189	No	Yes	m	12.7
13	m	14.5	149	No	No	I	m	14.4	235	No	No	m	13.5
14	m	15.3	545	No	No	I	m	15.5	526	No	No	m	15.5
15	m	15.7	994	No	No	I	m	16.0	1101	No	No	m	15.7
16	m	16.4	398	No	Yes	I	m	15.8	419	No	No	m	23.9
17	m	17.6	299	No	No	I	m	17.7	233	No	No	m	24.7
18	m	19.9	1135	Yes	No	I	m	22.6	853	Yes	No	m	25.8
19	m	21.2	235	No	No	I	m	21.5	240	No	No	m	27.2
20	f	29.0	182	No	Yes	I	f	29.6	252	No	No	f	29.3
21	f	34.9	148	No	No	I	f	32.1	228	No	No	f	33.0
22	m	36.4	408	No	No	I	m	35.3	528	No	No	m	35.6
23	m	36.6	220	No	No	II	m	35.1	244	No	Yes	m	35.7
24	m	37.5	169	No	Yes	I	m	38.0	122	No	Yes	m	37.8
25	m	37.7	260	No	No	II	m	38.3	543	Yes	Yes	m	41.4
26	m	38.7	326	No	Yes	I	m	46.1	423	No	No	m	43.2
27	m	46.7	505	Yes	No	II	m	55.3	415	No	No	m	51.0
28	f	52.3	990	Yes	No	II	f	49.1	626	Yes	Yes	f	50.4
29	f	63.5	180	No	No	II	f	46.2	186	Yes	No	f	51.7