Gait in children with arthrogryposis multiplex congenita

Marie Eriksson · Elena M. Gutierrez-Farewik ·
Eva Broström · Åsa Bartonek

Received: 14 October 2009 / Accepted: 21 December 2009 / Published online: 16 January 2010
© EPOS 2010

Abstract

Purpose Lower limb contractures and muscle weakness are common in children with arthrogryposis multiplex congenita (AMC). To enhance or facilitate ambulation, orthoses may be used. The aim of this study was to describe gait pattern among individuals wearing their habitual orthotic devices.

Methods Fifteen children with AMC, mean age 12.4 (4.3) years, with some lower limb involvement underwent 3-D gait analysis. Three groups were defined based on orthosis use; Group 1 used knee–ankle–foot orthoses with locked knee joints, Group 2 used ankle–foot orthoses or knee–ankle–foot orthoses with open knee joints and Group 3 used no orthoses.

Results The greatest trunk and pelvis movements in all planes and the greatest hip abduction were observed in Group 1, compared to Groups 2 and 3, as well as to the gait laboratory control group. Maximum hip extension was similar in Groups 1 and 2, but in Group 3, there was less hip extension and large deviations from the control data. Lower cadence and walking speed were observed in Group 1 than in Groups 2 and 3. The step length was similar in all

groups and also with respect to the gait laboratory reference values.

Conclusions Children with AMC were subdivided according to orthoses use. Kinematic data as recorded with 3-D gait analysis showed differences among the groups in trunk, pelvis and knee kinematics, and in cadence and walking speed. The step length was similar in all groups and to the gait laboratory reference values, which may be attributable to good hip extension strength in all participants.

Keywords Motion analysis · AMC · Amyoplasia · Ambulation

Introduction

Arthrogryposis multiplex congenita (AMC) can be described as a complex condition characterised by deformed joints with an intact sensory system [1, 2]. To be defined as AMC, there should be contractures in multiple joints in multiple body areas present at birth [1]. Arthrogryposis refers to a large heterogeneous group of conditions [3], with an incidence reported from 1/3,000 to 1/5,100 live births [1, 4]. The underlying cause of AMC is decreased foetal movements due to various factors, such as neuropathic and myopathic processes, abnormality of connective tissue, maternal diseases or impaired foetal or intrauterine vascularity. A correlation has been found between the early onset of insufficient movement and the severity of contractures [1].

AMC has been classified into three groups; disorders with mainly limb involvement (four limb, lower limb or upper limb), disorders with limb involvement associated

M. Eriksson · E. M. Gutierrez-Farewik · E. Broström ·
Å. Bartonek (✉)
Department of Women's and Children's Health,
Karolinska Institutet, MotorikLab Q2:07,
171 76 Stockholm, Sweden
e-mail: asa.bartonek@ki.se

E. M. Gutierrez-Farewik
KTH Mechanics, Royal Institute of Technology,
Stockholm, Sweden

with other organs, and disorders with limb involvement and central nervous dysfunction [1]. Various subtypes have also been distinguished, such as amyoplasia, distal arthrogryposis and Larsen syndrome. The most common subtype is amyoplasia, which represents one-third of all cases and is also usually referred to as classic arthrogryposis. In approximately half of the patients, a specific subtype diagnosis cannot be given [1].

In children with amyoplasia, hip deformities are common and range from soft tissue contractures to hip dislocation [3, 5]. Knee joint involvement has been reported in 70%, with flexion contractures being the most common, followed by extension contractures [5]. Foot deformities are frequently observed, with equinovarus adductus foot being the most common [5, 6]. Children with AMC frequently undergo orthopaedic surgery, often with multiple procedures [3, 7]. The goals for combining operative procedures are to avoid multiple immobilisation periods [8] and reduction of muscle strength [9].

The ability to walk depends on the extent of joint range of motion, in particular in the hips and knees, as well as in the foot, with possibility of plantigrade foot position. Muscle weakness in the lower extremities, primarily in the hip and knee extensor muscles, has also been reported to influence walking ability [10], and in children with amyoplasia, muscle weakness was considered to be more influential on walking ability than the severity of contractures [9]. The involvement of upper extremities with inability of hand and arm support has also been found to influence walking function [10, 11]. Most children with AMC can achieve functional ambulation and 85% have been reported as ambulators by the age of 5 years, but for efficient community ambulation, a wheelchair may be required [3, 12, 13].

To enhance or facilitate ambulation in children with AMC, orthoses may be used to compensate for muscle weakness and to support the lower extremities in an aligned position [8, 10, 13]. Orthoses are made of different materials and with or without knee-locking mechanisms [14]. The use of ankle carbon fibre spring orthoses has reportedly led to increased plantarflexor moment and stride length in children with AMC [15]. Shoe wedges have also been recommended to compensate for plantarflexion contractures [13]. In our clinical practice, effort is made to analyse the needs of orthoses in children with AMC and to evaluate gait outcome. With the exception of one study comparing two different types of orthoses in a group which included four children with AMC [15], no published description of gait in children with AMC has been found. The aim of this study was to describe gait pattern among individuals with AMC wearing their habitual orthotic devices.

Participants and methods

The study was approved by the local ethics committee and informed consent was obtained from the participants and their parents.

Participants

Of 27 children with AMC born between the years 1989 and 2003 and who were treated at the orthopaedic department at Karolinska University Hospital, 17 children fulfilled the criteria of some lower limb motor involvement and independent ambulation, and were invited to participate in a gait analysis study. Inclusion criteria were independent ambulation with or without orthoses and age between 4 and 18 years. Two children were excluded, one child due to an unclear diagnosis and one child with unilateral leg involvement who used a prosthesis, thus, 15 children (8 males, 7 females; mean age 12.4 [4.3] years [range 4.7–17.7]) participated in the study (Table 1). Six children had four-limb and nine had lower-limb involvement (Table 1).

Orthosis subgroups

Twelve of the 15 participants used orthoses (Table 1). The children were designated into subgroups with respect to orthosis use. Prescriptions of orthoses were based on the presence of muscle weakness, joint contractures or need of joint stabilisation according to the current orthotic programme. Group 1, represented by four participants, used knee–ankle–foot orthoses with locked knee joints (KAFO-L) (Fig. 1). Three participants had grade ≤ 3 in knee extensor muscle strength and one participant had grade 4 and bilateral knee flexion contractures of 25–30°. Two children had the knee locking mechanisms anteriorly and two posteriorly, depending on the child's hand function. Two participants had a carbon fibre ankle joint in combination with their KAFOs with locked knee joints (KAFO-L-C) (Fig. 2).

Group 2, with eight participants, used knee–ankle–foot orthoses with open knee joints (KAFO-O) or ankle–foot orthoses (AFO) of different types. Two participants had open knee joints with an extension stop and carbon fibre spring ankle joint (KAFO-O-C) (Fig. 3). In one child, this was required bilaterally to control knee hyperextension and knee valgus due to the lack of both anterior and posterior cruciate ligaments. In the other child, the control of knee valgus and knee hyperextension was indicated unilaterally, and an ankle–foot orthoses with a carbon fibre spring ankle joint (AFO-C) (Fig. 4) was used on the contralateral limb. Five participants used AFOs bilaterally and one unilaterally to stabilise the ankle joint. Four participants used an AFO-C with the aim to utilise the properties of the material to restore energy [15], one used a hinged (AFO-H) and one a solid (AFO-S).

Table 1 Distribution of patient characteristics, limb involvement, functional ambulation orthoses type and group according to orthosis use

Subject	Gender	Age (years)	Height (cm)	Weight (kg)	Limb involvement	Functional ambulation	Orthoses type (L/R)	Group
1	M	12.5	149	31.4	FL	III	KAFO-L-C	1
2	M	12.7	141	38.1	FL	III	KAFO-L-C	
3	M	13.0	165	80.0	LL	III	KAFO-L	
4	M	16.7	165	48.2	LL	III	KAFO-L	
5	M	4.7	112	19.7	LL	II	Shoe/AFO-C	2
6 ^a	F	6.5	131	27.6	LL	II	KAFO-O-C	
7 ^a	F	7.4	119	21.9	LL	II	AFO-C	
8 ^a	M	9.2	136	28.8	LL	II	AFO-C	
9 ^a	F	12.3	155	37.1	FL	II	AFO-H	
10	M	15.3	166	44.6	LL	II	AFO-C	
11	F	16.2	159	46.3	FL	III	KAFO-O-C/AFO-C	
12 ^a	M	17.7	170	55.8	FL	II	AFO-S	3
13	F	12.4	151	37.9	LL	I	Barefoot	
14	F	14.0	145	37.1	LL	I	Shoes with heel wedges	
15	F	13.9	159	38.6	FL	II	Shoes with heel height, unilateral	

R right; *L* left; *FL* four limb; *LL* lower limb; *KAFO* knee–ankle–foot orthoses; *L* locked knee joint; *C* carbon fibre spring ankle joint; *AFO* ankle–foot orthoses; *O* open knee joint with extension stop; *H* hinged; *S* solid

^a Gait analysis performed both with orthosis and barefoot

Fig. 1 Knee–ankle–foot orthoses with locked knee joint (KAFO-L)

All orthoses were made by the same orthotic company.

Group 3, with three participants, used no orthoses. Among the subjects in Group 3, one child preferred to walk barefoot and two children used shoes, of which one child had a unilateral lift due to leg length discrepancy and the other had heel wedges to compensate for plantarflexion contractures.

Fig. 2 Knee–ankle–foot orthoses with locked knee joint and a carbon fibre ankle joint (KAFO-L-C)

Fig. 3 Knee-ankle-foot orthoses with open knee joint and a carbon fibre ankle joint (KAFO-O-C)

Functional ambulation

Functional ambulation was assessed according to a five-level scale, which has been used previously in children with myelomeningocele [16, 17]. In Group 1, all participants were designated level III, i.e. household ambulators and wheelchair users for long indoor distances. In Group 2, one participant was level III and eight participants were level II, i.e. community ambulators who require a wheelchair for long distances outdoors only. In Group 3, one participant was level II and two participants were level I, i.e. community ambulators with no need for a wheelchair (Table 1).

Muscle strength

The strength of the lower limb muscles was tested manually according to a six-graded scale [18], with grade 0 indicating no muscle strength, grade 1 activity traces, grade 2 gravity-eliminated movement, grade 3 movement against gravity and grade 4 indicating movement against gravity

Fig. 4 Ankle-foot orthoses with a carbon fibre ankle joint (AFO-C)

with some manual resistance. Grade 5, indicating normal strength, was not given in this study. The distribution of muscle strength in the lower limbs according to Groups 1–3 is shown in Table 2.

Joint contractures

Passive range of motion was measured with a goniometer. Hip and knee flexion contractures were defined when measured $\geq 10^\circ$ and plantarflexion contractures as $>0^\circ$ from a neutral joint position. Two children had hip flexion contractures of $10\text{--}20^\circ$. Seven children had knee flexion contractures of $10\text{--}30^\circ$ and eight children, five in Group 2 and three in Group 3, had knee hyperextension of $10\text{--}20^\circ$. Seven children had restricted knee flexion of $20\text{--}110^\circ$ from the neutral position. Eight children had plantarflexion contractures of $10\text{--}20^\circ$ (Table 2).

Orthopaedic surgery

Fourteen participants had undergone orthopaedic surgery, of which 12 children underwent bony surgery and two children had only soft tissue surgery (Table 2).

Gait analysis

All children underwent 3-D gait analysis using an eight-camera motion analysis system (Vicon[®], Oxford, UK). The children were equipped with 34 reflective markers aligned

Table 2 Muscle strength, joint range of motion and previous orthopaedic surgery in all participants

Group	Child	Side ^b	Muscle strength grading according to a 0–5 scale ^a								Joint range of motion (°)					Orthopaedic surgery
			Hip			Knee		Ankle			Hip ^c		Knee ^d		Ankle ^e	
			Flex L/R	Ext L/R	Abd L/R	Flex L/R	Ext L/R	Dors L/R	Plant L/R	Ext L/R		Flex L/R	Ext L/R	Dors L/R		
1	1	L	4/3	3/4	4/4	4/3	2/3	0/0	0/0	0/0		130/50	–15/0	–20/–20	Bi hip, bi ankle	
	2	R	3/3	4/4	4/4	4/4	3/3	0/0	0/0	–10/–20	60/60	–30/–25	–20/–15	Bi knee, bi ankle		
	3	R	3/3	4/3	4/3	2/3	2/2	0/0	0/0	0/0	100/110	0/–20	–15/–15	Bi knee, bi ankle		
	4	L	4/4	4/4	3/4	3/4	4/4	0/0	0/0	0/0	100/100	–30/–25	10/0	Bi knee ^f , bi ankle		
2	5	R	4/4	4/4	4/3	4/4	4/4	4/3	4/3	0/0	130/130	+20/–10	20/–15	R ankle ^f		
	6	R	4/4	4/4	4/4	4/4	4/4	4/4	4/4	0/0	150/150	+20/+20	0/10	–		
	7	L	4/4	4/4	4/4	4/4	4/4	4/4	3/4	0/0	130/140	+15/+15	0/0	Bi hip, L ankle		
	8	R	4/4	4/4	4/4	4/4	4/4	4/4	3/3	0/0	150/150	+10/+10	15/20	Bi ankle		
	9	L	4/4	4/4	4/4	4/4	4/4	0/4	0/3	0/0	140/140	–10/+10	–20/5	Bi ankle		
	10	R	4/4	4/4	3/3	3/2	4/4	0/0	0/0	0/0	60/60	–10/–10	–20/–20	L hip, bi ankle		
	11	L	4/4	4/4	4/4	4/4	4/4	2/2	2/2	0/0	40/110	+5/0	0/0	L hip, bi ankle		
	12	L	4/4	4/4	4/4	4/4	4/4	4/4	4/4	0/0	140/140	0/0	–20/0	Bi ankle		
3	13	R	4/4	4/4	4/4	4/4	4/4	4/4	4/4	0/0	120/20	+5/+10	5/10	Bi hip		
	14	L	4/4	4/4	3/3	4/4	4/4	4/4	4/4	–20/–15	140/140	+10/+15	–15/–10	Bi hip, bi ankle		
	15	R	4/4	4/4	4/4	4/4	4/4	4/4	4/4	0/0	155/155	+15/+15	15/10	R hip ^f		

Flex flexion; *Ext* extension; *Abd* abduction; *Dors* dorsiflexion; *Plant* plantarflexion; *L* left; *R* right; *bi* bilaterally

^a Muscle strength grade <4 was not applied in this study

^b Selected side for data analysis

^c 0 indicates neutral joint position and – indicates <0

^d Flexion indicates joint angle from 0 to full knee flexion range (140°), extension indicates <0 (–) and >0 (+)

^e 0 indicates neutral joint position and – indicates <0

^f Soft tissue surgery

with anatomical landmarks on the head, trunk and pelvis, and bilaterally on the arms, thighs, shanks and feet. The markers were placed by the same examiner (ÅB). The lower body was modelled according to the Newington model [19] and the upper body was modelled as the thorax, upper and lower arms, hands and head according to the Plug-in Gait model (Vicon). In the children who wore orthoses, the markers were placed as near as possible to the correct anatomical position. The subjects were asked to walk at a self-selected comfortable pace along a 10-m walkway until complete information from several gait cycles for each side was collected. In five of the six children who were able to walk without orthoses, gait analysis was performed barefoot.

Data analysis

Three kinematic gait cycles were generated for each subject. The following kinematic parameters were obtained from each gait cycle and averaged for each side to describe gait: range of lateral trunk sway, average trunk tilt, range of

trunk rotation, pelvic elevation range, average pelvic tilt, pelvic rotation range, maximum hip abduction, maximum hip flexion and extension, hip rotation at initial contact, knee flexion at initial contact and in mid stance, maximum knee flexion, knee flexion/extension range and average foot progression in stance. For Group 3, the maximum dorsiflexion and plantarflexion were also analysed. Time and distance parameters were analysed, wherein velocity, step length and stride length were normalised to the leg length.

Statistical analysis

The Wilcoxon signed ranks test was used to test for differences between the left and right sides within each of the groups and between gait during barefoot walking and with orthoses in five participants in Group 2 (Table 1). The Kruskal–Wallis test was used to compare values between the three groups. All statistical analyses were carried out using commercially available software (SPSS version 16.0). A *P*-value of ≤0.05 was considered to be statistically significant.

Results

There were no statistical significant differences between the right and left sides in any parameter within any group. For data presentation and subsequent between-group data analysis, the limb with less muscle strength and/or greater contractures was selected. When there was no difference between the limbs, the right side was selected (Table 2).

The results are presented with respect to the orthosis groups. Reference kinematic data is illustrated by the gait laboratory control group, consisting of 23 healthy children aged 5–14 years.

Trunk kinematics

In Group 1, anterior-posterior trunk tilt movements tended towards posterior in stance and swing, which was also slightly pronounced in Group 2. In Groups 2 and 3, the participants were relatively similar in all planes except one participant in Group 3 who showed greater posterior trunk tilt of approximately 11° during the entire gait cycle. Between the groups, the trunk lateral sway range ($P = 0.015$) and trunk rotation range ($P = 0.007$) differed significantly (Fig. 5, Table 3).

Pelvis kinematics

In Group 1, internal rotation was more pronounced in stance and external rotation in swing. In all groups, there were various pelvic obliquity movements among the participants, with pelvic lift in swing frequently observed in Group 1. The mean anterior/posterior pelvic tilt values were greatest in Group 3 (Table 3) and movements varied among participants in all groups. Pelvis rotation range differed significantly between the groups ($P = 0.021$) (Fig. 5, Table 3).

Hip kinematics

Group 1 showed an average of 2–3 times greater maximum hip abduction than Groups 2 and 3 (Table 3), although there were variations among participants in Groups 1 and 2. The maximum hip extension was similar in Groups 1 and 2, reaching almost neutral position, with reasonably similar movements among participants. Group 3 had less hip extension than Groups 1 and 2 (Table 3). The maximum hip flexion was similar in all groups (Table 3), with relatively similar movements among the participants in all groups. The mean hip rotation at initial contact was similar in the groups (Table 3), but large deviations among participants were observed in Group 1 (Fig. 6).

Knee kinematics

The maximum knee flexion in swing as well as the mean flexion/extension range were lowest in Group 1 ($P = 0.048$ and 0.015 , respectively) (Table 3), as a consequence of the locked orthotic knee joints. Despite the locked orthoses in Group 1, corresponding to the knee flexion contractures, there was greater knee flexion at initial contact and greater knee flexion at mid stance compared to Groups 2 and 3, with variation among Group 1 (Table 3). In Group 2, the knee flexion/extension movements were similar among the participants. Reduced knee flexion during swing was observed in one participant in Group 3 due to restricted knee flexion motion (Fig. 6).

Foot kinematics

Dorsiflexion and plantarflexion for Groups 1 and 2 were not analysed due to various orthoses and footwear conditions. Group 1 showed large variation among the participants, with both large internal as well as external foot progression (Fig. 6, Table 3).

Time and distance parameters

Lower cadence and lower walking speed were observed in Group 1 than in Groups 2 and 3 ($P = 0.019$ and 0.033 , respectively). The step length and stride length were similar in the groups ($P = 0.341$ and 0.460 , respectively). Group 1 showed somewhat greater step width compared to Groups 2 and 3, though the difference was not statistically significant ($P = 0.072$) (Table 3).

Observations with respect to the gait laboratory kinematic control data and time and distance parameters reference values

Upon visual comparison of the AMC groups' data with respect to the gait laboratory control data, the greatest deviations in trunk movements were seen in Group 1 in all planes. In the pelvis, great deviations were observed in Group 1 in all movements, in Group 2 in increased pelvic obliquity in swing and in both Groups 2 and 3 in increased anterior pelvic tilt. At the hip, obvious discrepancy was observed in Group 1 as increased abduction and hip rotation, and in Group 3 as less maximum hip extension. At the knee, discrepancy was observed in Group 1 in knee flexion/extension movements during the entire gait cycle, and in Group 3 as less flexion in swing in one participant. At the foot, discrepancy was observed in Group 1 in variation

Fig. 5 Illustrations of movements in the trunk and pelvis in all planes for Groups 1–3. The *shaded field* represents the mean ± 1 standard deviation (SD) of the gait laboratory control group, and each line represents an individual in each group, respectively

Table 3 Mean (standard deviation [SD]) of the trunk, pelvis and lower limb joint angles, cadence, step length, stride length, walking speed and step width in Groups 1–3

	Group 1 (<i>n</i> = 4) Mean (SD)	Group 2 (<i>n</i> = 8) Mean (SD)	Group 3 (<i>n</i> = 3) Mean (SD)	<i>P</i> -value ^k
Trunk (°)				
Lateral sway (range)	21.4 (6.9)	6.5 (2.8)	5.2 (2.3)	0.015
Tilt ant/post ^a (average)	−1.9 (5.0)	1.9 (4.4)	3.5 (6.6)	0.419
Rotation (range)	22.6 (12.5)	8.7 (2.1)	6.5 (1.5)	0.007
Pelvis				
Elevation (range)	13.5 (6.2)	10.0 (5.1)	10.9 (4.1)	0.322
Tilt ant/post ^b (average)	18.8 (5.8)	15.6 (6.4)	29.5 (11.3)	0.126
Rotation (range)	34.7 (11.2)	13.9 (5.6)	22.5 (10.9)	0.021
Hip				
Abduction ^c (max.)	−18.1 (9.9)	−8.7 (5.7)	−5.8 (3.2)	0.109
Extension ^d (max.)	−4.4 (8.0)	−1.5 (8.4)	17.2 (4.1)	0.055
Flexion (max. ^e)	44.0 (11.3)	41.6 (11.5)	52.9 (9.4)	0.347
Rotation ^f (IC ^g)	−7.0 (34.3)	−7.8 (10.1)	−9.4 (13.5)	0.989
Knee				
Flexion (IC ^g)	17.9 (7.4)	6.9 (6.5)	3.0 (7.7)	0.052
Flexion (MS ^h)	15.8 (9.7)	5.7 (9.4)	9.2 (4.8)	0.205
Flexion (max. ^e)	19.3 (7.4)	55.1 (9.2)	40.8 (27.3)	0.048
Flex/ext (range)	4.7 (2.3)	51.8 (11.6)	39.4 (23.2)	0.015
Ankle				
Dorsiflexion (max.)	–	–	7.4 (5.0)	–
Plantarflexion ⁱ (max.)	–	–	−17.6 (4.2)	–
Foot progression ^f (average ^j)	−0.7 (24.8)	−7.4 (7.1)	−8.2 (5.0)	0.853
Time and distance parameters				
Cadence (steps/min)	87 (6)	120 (16)	128 (5)	0.019
Step length/leg length	0.71 (0.05)	0.81 (0.14)	0.74 (0.03)	0.341
Stride length/leg length	1.46 (0.10)	1.63 (0.26)	1.43 (0.03)	0.46
Walking speed/leg length (s ^{−1})	1.06 (0.13)	1.65 (0.45)	1.53 (0.06)	0.033
Step width (m)	0.24 (0.07)	0.16 (0.05)	0.12 (0.04)	0.072

^a + = posterior^b + = anterior^c − = abduction^d − = extension^e Max in swing phase^f − = external^g Initial contact^h Mid stanceⁱ − = plantarflexion^j Stance^k Comparison between AMC groups

among the participants and in Group 2 in increased internal foot progression in swing (Figs. 5 and 6).

Compared to the control group's average (standard deviation) time and distances parameters, cadence 130 (14) steps/min, step length/leg length 0.79 (0.06), stride length/leg length 1.59 (0.12), walking speed/leg length

1.72 (0.26) s^{−1} and step width 0.14 (0.02) m, it was notable that Groups 2 and 3 had similar cadence, all groups had step length relatively close to the control group, Group 2 had longer stride length and very similar walking speed, and Group 1 had almost twice the step width.

Fig. 6 Illustrations of movements of the hip in all planes, and in the knee and foot in the sagittal plane for Groups 1–3. The *shaded field* represents the mean ± 1 SD of the gait laboratory control group and each line represents an individual in each group, respectively

Time and distance parameters during barefoot walking and with orthoses

The results were compared between walking barefoot and with orthoses in five participants in Group 2. With

orthoses, the mean cadence was slightly lower, the mean step length increased by approximately 0.09 m, the mean stride length increased by 0.17 m and mean walking speed increased by approximately 0.25 m/s, but there were no significant differences (Table 4).

Table 4 Mean (SD) of cadence, step length, stride length and walking speed in five patients in Group 2 comparing barefoot and walking with orthoses

<i>N</i> = 5	Barefoot Mean (SD)	Orthoses Mean (SD)	<i>P</i> -value
Cadence (steps/min)	132 (8)	127 (21)	0.5
Step length (m)	0.49 (0.06)	0.58 (0.09)	0.08
Stride length (m)	1.00 (0.17)	1.17 (0.17)	0.08
Walking speed (m/s)	1.09 (0.21)	1.34 (0.27)	0.138

Discussion

By subdividing the participants according to the orthoses they had been prescribed, we identified different walking patterns. This subject population is heterogeneous, and orthosis prescription, while not an obvious criterion for group definition, was indicative to a large extent of subjects' physical properties and body function. The participants in Group 1 who used KAFOs with locked knee joints displayed more extensive trunk and pelvic movements, hip abduction/adduction as well as hip rotation movements compared to the other groups, including those three participants who used no orthoses. A typically wide-based walking pattern in children with AMC wearing KAFOs has been described [20]. In Group 1, wider steps and more abducted hip movements were found compared to the other two groups, which may be interpreted as increasing their support base to improve balance, but also as compensatory mechanisms for the extended knees.

Hip flexion and extension movements in all groups were similar to the gait laboratory control group, but less hip extension in late stance was seen in all groups. In Group 3, who did not require orthoses, two children had passive hip extension range of motion to the neutral position, but did not utilise their full hip extension during stance, possibly due to previous hip deformities, and one child had hip flexion contractures.

The step length was similar in all groups and to the gait laboratory reference value, which may be attributable to good hip extension strength. The importance of hip extension strength grade 4 or better for functional ambulation in patients with AMC has been reported [10] and good correlation has been found between hip muscle strength and motor function in patients with amyoplasia [9]. Even if it is not possible to measure the muscle strength over the entire range of motion, it is important to grade the resistance throughout the available arc of motion, since children with AMC may be strong in the midrange [13].

All of the children in this study were able to perform movements against gravity with or without manual resistance in hip flexion, extension and abduction, and most of

them in knee extension and flexion. KAFOs with locked knee joints have been recommended if knee extensor weakness or a knee flexion contracture is present [5, 10]. KAFOs with open knee joints have frequently been used by children with myelomeningocele to stabilise the knee joint in the frontal and transverse planes [21]. In children with AMC, both flexion contractures [10] and hyperextension have frequently been reported, wherein flexion contractures, in contrast to hyperextension, have a negative influence on walking ability [5, 22]. In our group, five knee joints reached a neutral position, ten had knee flexion contractures and 15 knees could hyperextend. In one child, knee hyperextension was excessive, which made an extension stop necessary. The thigh segment also improved the control of the knee in the frontal and transverse planes.

In AMC, complete correction of clubfoot has been described as difficult to achieve [23] and residual stiffness has been reported [6], which can cause ambulatory difficulties [6, 10]. In a recently published long-term follow-up study [24], all independent or community walkers were reported to have plantigrade feet, despite foot deformities at birth. In children with joint contractures, improved postural alignment of the body segments can be obtained by adding wedges under the heels [13]. Such wedges were used in one child in Group 3 (subject 14). Six of eight participants in Group 2 used an energy-restoring carbon fibre ankle joint in their orthoses. In four of six participants, the stride length was longer with respect to the control group data, which is in accordance with previous results [15]. The greater step length and walking speed observed in Group 2 compared to Group 3 may also be attributable to the use of ankle carbon fibre springs.

Six of eight children in Group 2 were able to walk independently without orthoses, of which one child could only do so with difficulty. The results of the five participants who performed gait analysis both barefoot and with orthoses indicate that time and distance parameters improved during walking with orthoses; however, no significant differences could be found in such a small number.

All participants used their orthoses regularly throughout the day, which indicates high acceptance of the orthoses. In Group 1, all children required orthoses to achieve the ability to walk. With orthoses, they were able to walk indoors and even short distances outdoors. The children who used KAFO-L were able to lock and unlock the knee joints independently, and were also able to change their position from sitting to standing. Three children required help putting on the orthoses due to impaired hand function. Limited ambulation due to poor protective responses of the upper extremity has been reported [13] and that children wearing KAFOs often require walkers [20]. In this study group, only one child

occasionally used a walking aid, in the school yard for safety reasons.

The ambulatory activity level in youths with AMC has been reported as being lower compared to a control group [25]. In our study, all four children in Group 1 were household ambulators and of the total of seven children who used a wheelchair, six also had a powered wheelchair, four of whom had impaired hand function. Future studies of energy consumption can provide more information about the physical effort during walking in children with AMC.

Conclusion

Gait pattern in children with arthrogryposis multiplex congenita (AMC) was recorded with their orthoses, ranging from locked knee joints to ankle foot orthoses and shoes only. We have shown differences among the groups in the trunk, pelvis and knee kinematics, and in cadence and walking speed. In the children requiring locked knee joints, the greatest trunk and pelvis movements and the lowest knee flexion were observed, as well as the lowest cadence and slowest walking speed.

The step length was similar in all groups and to the gait laboratory reference values, which may be attributable to good hip extension strength in all participants. Comparison between barefoot and orthotic condition was performed in only five participants, and indicated improved stride and temporal parameters, though the small participant number precludes conclusions on orthosis benefit.

Acknowledgments We would like to thank the children and their parents for participating in the study. This study was supported by the Norrbacka-Eugenia Foundation.

References

- Hall JG (1997) Arthrogryposis multiplex congenita: etiology, genetics, classification, diagnostic approach, and general aspects. *J Pediatr Orthop B* 6:159–166
- Thompson GH, Bilenger RM (1985) Comprehensive management of arthrogryposis multiplex congenita. *Clin Orthop Relat Res* 194:6–14
- Bevan WP, Hall JG, Bamshad M, Staheli LT, Jaffe KM, Song K (2007) Arthrogryposis multiplex congenita (amyoplasia): an orthopaedic perspective. *J Pediatr Orthop* 27:594–600
- Darin N, Kimber E, Kroksmark AK, Tulinius M (2002) Multiple congenital contractures: birth prevalence, etiology, and outcome. *J Pediatr* 140:61–67
- Staheli LT (1998) Lower extremity management. In: Staheli LT, Hall JG, Jaffe KM, Paholke DO (eds) *Arthrogryposis: a text atlas*. Cambridge University Press, Cambridge, pp 55–73
- Guidera KJ, Drennan JC (1985) Foot and ankle deformities in arthrogryposis multiplex congenita. *Clin Orthop Relat Res* 194:93–98
- Carlson WO, Speck GJ, Vicari V, Wenger DR (1985) Arthrogryposis multiplex congenita. A long-term follow-up study. *Clin Orthop Relat Res* 194:115–123
- Staheli LT (1998) Orthopedic management principles. In: Staheli LT, Hall JG, Jaffe KM, Paholke DO (eds) *Arthrogryposis: a text atlas*. Cambridge University Press, Cambridge, pp 27–43
- Kroksmark AK, Kimber E, Jerre R, Beckung E, Tulinius M (2006) Muscle involvement and motor function in amyoplasia. *Am J Med Genet A* 140:1757–1767
- Hoffer MM, Swank S, Eastman F, Clark D, Teitge R (1983) Ambulation in severe arthrogryposis. *J Pediatr Orthop* 3:293–296
- Hahn G (1985) Arthrogryposis. Pediatric review and rehabilitative aspects. *Clin Orthop Relat Res* 140:104–114
- Sells JM, Jaffe KM, Hall JG (1996) Amyoplasia, the most common type of arthrogryposis: the potential for good outcome. *Pediatrics* 97:225–231
- Donohoe M (2006) Arthrogryposis multiplex congenita. In: Campbell SK, Vander Linden DW, Palisano RJ (eds) *Physical therapy for children*, 3rd edn. Saunders Elsevier Inc., St. Louis, pp 381–400
- Florence J (1977) The orthotic management of arthrogryposis. *Prosthet Orthot Int* 1:111–113
- Bartonek Å, Eriksson M, Gutierrez-Farewik EM (2007) Effects of carbon fibre spring orthoses on gait in ambulatory children with motor disorders and plantarflexor weakness. *Dev Med Child Neurol* 49:615–620
- Bartonek Å, Saraste H (2001) Factors influencing ambulation in myelomeningocele: a cross-sectional study. *Dev Med Child Neurol* 43:253–260
- Danielsson AJ, Bartonek Å, Levey E, McHale K, Sponseller P, Saraste H (2008) Associations between orthopaedic findings, ambulation and health-related quality of life in children with myelomeningocele. *J Child Orthop* 2:45–54
- Hislop HJ (2007) Daniels and Worthingham's muscle testing: techniques of manual examination, 8th edn. Saunders Elsevier Inc., St. Louis
- Davis RB, Öunpuu S, Tyburski D, Gage JR (1991) A gait analysis data collection and reduction technique. *Hum Mov Sci* 10:575–587
- Gaubert CS, Chaplin DL, Jaffe KM (1998) Physical and occupational therapy. In: Staheli LT, Hall JG, Jaffe KM, Paholke DO (eds) *Arthrogryposis: a text atlas*. Cambridge University Press, Cambridge, pp 87–113
- Gutierrez EM, Bartonek Å, Haglund-Åkerlind Y, Saraste H (2003) Characteristic gait kinematics in persons with lumbosacral myelomeningocele. *Gait Posture* 18:170–177
- Murray C, Fixsen JA (1997) Management of knee deformity in classical arthrogryposis multiplex congenita (amyoplasia congenita). *J Pediatr Orthop B* 6:186–191
- Ponseti IV (1996) Congenital clubfoot. Fundamentals of treatment. Oxford University Press Inc., New York
- Fassier A, Wicart P, Dubosset J, Serigne R (2009) Arthrogryposis multiplex congenita. Long-term follow-up from birth until skeletal maturity. *J Child Orthop* 3:383–390. doi:10.1007/s11832-009-0187-4
- Dillon ER, Bjornson KF, Jaffe KM, Hall JG, Song K (2009) Ambulatory activity in youth with arthrogryposis: a cohort study. *J Pediatr Orthop* 29:214–217