



## Research paper

# Hip abduction and supported standing affect the ranges of hips extension in spinal muscular atrophy patients

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### ABSTRACT

**Introduction:** Recommendations for management of spinal muscular atrophy (SMA) do not contain detailed information about the position of lower limbs during support standing. It has been observed that during the measurement of the range of extension in the hip joint (HE) in SMA patients, the examined limb was often naturally abducted.

**Aim:** The main aim of the study was to compare the values of HE in the sagittal plane and in abduction, and to assess the correlation between the duration of supported standing and HE in SMA patients.

**Material and methods:** The study group consisted of 75 SMA individuals aged 2–22 years and control group consisted of 202 healthy participants. The measurements were performed with the Rippstein plurimeter and goniometer.

**Results and discussion:** Range of HE in SMA patients was larger in abduction than in the sagittal plane. A correlation was noted between the duration of supported standing and HE.

**Conclusions:** Supported standing with hip joint abduction should be used in SMA patients. The obtained results broaden the knowledge about the biomechanics of hip joints in SMA patients.

## 1. INTRODUCTION

Spinal muscular atrophy (SMA) is a rare autosomal recessive neuromuscular disease characterised by the degeneration of motor neurons.<sup>1,2</sup> Patient care is problematic due to the progression of muscle weakness, breathing disorders, bone deformities, joint contractures or problems with chewing and swallowing.<sup>2–11</sup>

The need for systematic physical therapy including, i.a., stretching and support standing, is indicated in the standards of care for SMA.<sup>5,10</sup>

A serious problem in management of SMA is constituted by movement limitations, subluxations and dislocations of hip joints.<sup>3,5,7,8,11–15</sup> Both incomplete mobility and hip joint dislocation hinder everyday functioning of patients and limit possibilities of supported standing, which has a positive effect on the organism. Axial loading decreases the risk of scoliosis,<sup>5</sup> forces bones to grow and constitutes training for the cardiovascular and respiratory systems. It also limits the occurrence of contractures and spinal deformities in patients with neuromuscular diseases who lost the ability to walk.<sup>16</sup> Moreover, due to new possibilities of pharmacological treatment for SMA patients,<sup>17–19</sup> the standing program should be treated as a manner of maintaining a standing position or preparing for walking.

The recommendations for rehabilitation specify the time and frequency of supported standing for SMA patients.<sup>10</sup> Unfortunately, there are no detailed guidelines regarding the position in which patients should be positioned. It is recommended that hip joints in children should be positioned in abduction, but studies were conducted in children with other diseases, especially with cerebral palsy.<sup>20,21</sup> Due to the lack of detailed guidelines for SMA patients, the systems with parallel lower limbs alignment are often used for support standing.

One of the factors determining a proper body position while standing is an appropriate range of extension in hip joints (HE). The authors' experience shows that numerous SMA patients reported pain in the spinal area and lower limbs which occurred during supported standing. It was also noticed that one or both lower limbs positioned in extension in the hip joints in a supine position were often naturally abducted (Figure 1A) and the range of HE in the sagittal plane was more limited than in an abducted position (Figure 1B). Both the position and the ranges of motion in hip joints affect an alignment of the body while standing (Figure 1C).

Therefore, there occurs a question regarding the position in which SMA patients should practise standing, particularly taking into account the fact that many patients experience hip joint dislocation while growing.



**Figure 1.** Influence of range of motion of the hip on support standing position: A preferred abduction in extension in the right hip joint in the boy with SMA type 2 (1A); Flexion contracture in the right hip joint in the sagittal plane (1B); Asymmetric position of lower limbs and an improper body position during supported standing (1C).

## 2. AIM

The main aims of the study were: (1) to compare the values of the range of passive HE in the sagittal plane and in abduction, if it occurred, in SMA patients; (2) to gather information regarding the duration and quality of supported standing; (3) to determine the correlation between the duration of supported standing and the ranges of HE. Additionally the values of the range of passive HE in the sagittal plane obtained in SMA individuals and the control group were compared to determine the frequency of the occurrence of movement limitation in the hip joints in SMA group.

## 3. MATERIAL AND METHODS

### 3.1. Participants

Individuals aged 2–22 years with SMA type 1 (SMA1), 2 (SMA2) and 3 (SMA3) confirmed in a genetic examination were qualified to the study group. Exclusion criteria included spinal or hip joint surgery, constant use of respirator and difficulties in taking a supine position due to a severe spinal deformity.

The control group included healthy children and youth aged 2–18 who attended posture examination.

The study was conducted during the Conference of the SMA Foundation Poland associating SMA patients and their families as well as during individual consultations in a physiotherapy centre.

### 3.2. Study protocol

The examination performed by experienced physiotherapists involved measuring the ranges of extension in hip joints as well as gathering information on the manner and duration of supported standing, the existence of scoliosis and dislocations in hip joints. The examination of hip joint extension was performed with Rippstein plurimeter on a patient in the standard supine position for testing the length of the hip flexors.<sup>22</sup> The plurimeter was zeroed in a position parallel to the surface and was placed on a thigh above the knee joint at the base of patella. The values of extension below the level were marked with '+,' while the values above the level referring to a bend in the hip joint were marked with '-.' Beforehand, the reliability of the measurements of the range of hip extension performed with the plurimeter was assessed in a group of SMA patients and an excellent level of intraobserver and interobserver reliability was revealed.<sup>23</sup>

In each patient, HE in the left (LHE) and right hip (RHE) in the sagittal plane, as well as in an individual preferred abduction (if it occurred) (ALHE, ARHE) was measured (Figure 2A). The abduction angle was measured with a goniometer (Figure 2B).



**Figure 2.** Measurements of range of motion of the hip: Measurement of HE in the sagittal plane with a plurimeter (A); Preferred abduction measured with a goniometer (B).

According to the methodology of the previous research, the limitation of extension by a minimum of 5° was perceived as a flexion contracture.<sup>3</sup> A mean value of extension in the control group was adopted as an appropriate range.

Afterwards, differences between the LHE and RHE values obtained in the sagittal plane (HED) and in abduction (AHED) were calculated in order to define a potential cause-and-effect interdependence between uneven ranges of extension in both hips and the occurrence of scoliosis in SMA individuals.

After verifying normal distribution with the Shapiro–Wilk test, the Kruskal–Wallis, Mann–Whitney and Bonferroni tests were applied. The correlation between the duration of supported standing and ranges of extension in hip joints was analysed with the use of Spearman correlation coefficient ( $r$ ). The level of significance was set at 0.05.

## 4. RESULTS

### 4.1. General information about participants

The study group consisted of 75 individuals with SMA aged 2–22. Children and youth constituted the majority of participants with SMA; however, there were 2 adults in the SMA group. In 22 (29.3%) SMA patients a subluxation or dislocation had been diagnosed.

The control group included 202 healthy individuals aged 2–18. Information regarding age, sex, body mass and height in SMA, SMA1, SMA2, SMA3 and the control group is included in Table 1.

### 4.2. The values of extension in hip joints

The mean values of the range of HE in SMA, SMA1, SMA2, SMA3 and control groups are shown in the Table 2.

**Table 1. The number, age, sex, weight and height of the participants in SMA, SMA1, SMA2, SMA3 and the control group.**

Group	Age, years mean ± SD	Weight, kg mean ± SD	Height, cm mean ± SD	Number of patients by sex		Total
				Females	Males	
SMA	6.1 ± 3.9	19.3 ± 9.6	115.1 ± 18.6	32	43	75
SMA1	6.3 ± 4.2	18.7 ± 7.0	117.0 ± 17.6	7	11	18
SMA2	6.4 ± 3.7	20.1 ± 9.7	117.0 ± 17.7	16	23	39
SMA3	5.1 ± 3.9	18.3 ± 12.0	109.2 ± 21.2	9	9	18
Control	6.8 ± 3.0	25.3 ± 11.4	124.6 ± 19.5	94	108	202

**Table 2. Values of the HE in the sagittal plane and abduction in SMA, SMA1, SMA2 and SMA3 groups, and in the sagittal plane in the control group, in degrees.**

Group / HE measurement	Mean	SD	Maximum	Minimum	Median
<b>SMA</b>					
LHE	-10.9	23.5	28.0	-62.0	-9.00
RHE	-10.4	22.9	26.0	-65.0	-8.00
ALHE	-2.8	20.4	26.0	-60.0	0.00
ARHE	-2.4	21.5	28.0	-58.0	0.00
<b>SMA1</b>					
LHE	-22.1	16.7	4.0	-60.0	-19.0
RHE	-21.7	14.1	0.0	-46.0	-21.0
ALHE	-10.4	17.6	16.0	-60.0	-5.0
ARHE	-9.0	17.2	14.0	-46.0	-5.0
<b>SMA2</b>					
LHE	-16.0	22.8	20.0	-62.0	-10.0
RHE	-15.1	22.9	20.0	-65.0	-10.0
ALHE	-6.4	20.0	24.0	-46.0	-5.0
ARHE	-6.8	22.2	24.0	-56.0	-2.0
<b>SMA3</b>					
LHE	11.4	15.7	28.0	-44.0	15.0
RHE	11.0	15.3	26.0	-42.0	14.0
ALHE	10.4	17.8	22.0	-44.0	17.0
ARHE	11.7	17.1	28.0	-42.0	18.0
<b>Control</b>					
LHE	20.7	4.9	36.0	0.0	20.0
RHE	19.9	5.4	38.0	0.0	20.0

In SMA group, the values of hip extension depended on the position of a lower limb in the coronal plane. In 60 participants (80%) lower limbs were naturally abducted during the measurement of the range of extension. In 15 study participants, no tendency to abduction was noted. These were mainly children aged 2–5 without limitations in the range of motion in hip joints or walking patients with SMA3.

A mean abduction angle in the left hip joint was 16.8° (SD 5.4°), while in the right joint it was 17.5° (SD 6.0°). Values of abduction observed in the SMA1 (left hip 17.9°, SD 5.4°; right hip 19.2°, SD 6.4°) and SMA2 (left hip 17.1°, SD 5.6°; right hip 17.9°, SD 5.7°) groups were higher than in the SMA3 group (left hip 13.0°, SD 1.8°; right hip 12.4°, SD 4.8°). In the majority of SMA participants (49 patients), the values of abduction in both hip joints ranged from 10° to 24°. In 4 participants, the preferred abduction in the hip joint was 30° and more.

Mean ranges of LHE and RHE in SMA group were significantly smaller than ALHE and ARHE ( $P < 0.05$ ).

The largest ranges of HE were noted in the SMA3 group, while the most limited ones in the SMA1 group (Table 2). Significant differences were noted between SMA1 and SMA3 ( $P < 0.01$ ) and between SMA2 and SMA3 groups ( $P < 0.01$ ) both in the sagittal plane and in abducted position. The differences between SMA1 and SMA2 were not significant.

The ranges of HE in SMA individuals were significantly limited compared to the control group ( $P = 0.000$ ). In 65 individuals with SMA (86%), a flexion contracture in both hip joints in the sagittal plane was noted. Among 11 patients without a flexion contracture there were 2 individuals with SMA2 and 8 individuals with SMA3.

The difference between LHE and RHE was larger than 4° in 48% of SMA participants in the sagittal plane (HED) and in 47% in abduction (AHED).

#### 4.3. The values of extension in hip joints with regard to age in SMA individuals

The ranges of LHE and RHE in particular age groups differed significantly. The children from the age groups of 2–3 year-olds and 4–6 year-olds achieved better ranges of HE than the participants aged more than 6 ( $P < 0.001$ ). No significant difference between the groups of 2–3-year-olds and 4–6-year-olds was noted ( $P = 0.158$ ).

The ranges of ALHE and ARHE also differed depending on age. The group of the participants aged more than 6 had significantly larger contractures than the group of 2–3-year-olds ( $P < 0.001$ ) and 4–6-year-olds ( $P < 0.001$ ). No significant difference between the groups of 1–3-year-olds and 4–6-year-olds was noted ( $P = 0.191$ ).

#### 4.4. Correlation between the range of HE and duration of supported standing in individuals with SMA

Different forms of standing were confirmed by 61 SMA participants, including 9 individuals with SMA1, 35 with SMA2 and 17 with SMA3. In this group, 2 SMA3 patients did not walk, but practised standing, 10 SMA3 individuals

walked with support and additional supported standing was applied, while 5 participants with SMA3 walked independently and did not undergo other forms of supported standing. Due to considerable limitations in hip joints, pain occurring during supported standing or poor physical state, 14 participants did not practise standing. Mean duration of supported standing was  $49.5 \pm 22.2$  min/day. A significant correlation was noted between the duration of supported standing and the range of HE in a neutral position ( $r = -0.312$ ,  $P = 0.001$ ) and a preferred position ( $r = -0.259$ ,  $P = 0.008$ ). In the case of individuals who were standing for a longer time, milder contractures in hip joints were noted. In 46 patients who underwent supported standing, standing frames or other supported standing systems with a parallel position of lower limbs were applied. In the case of 9 patients, supported standing in abduction was implemented. In 19 out of 22 individuals, whose subluxation or dislocation of a hip joint was confirmed radiologically, supported standing was applied.

#### 4.4. Differences in the ranges of extension in hip joints and scoliosis in SMA individuals

Thirty-eight study participants (53%) had scoliosis. Differences between the ranges of HE were larger in individuals with scoliosis than in individuals without scoliosis. The difference occurred to be significant in the case of AHED ( $P = 0.005$ ). A similar tendency was observed during the analysis of the values of HED ( $P = 0.053$ ).

## 5. DISCUSSION

Patients with SMA and their families frequently ask for the indication and the quality of supported standing. Implementation of the standing program is often difficult, both due to the lack of detailed guidelines and because of contractures in lower limb joints, spinal deformities as well as frequent subluxations or dislocations in hip joints. For this reason, these issues were the focus of our research.

The results obtained in our study confirm the findings of other researchers who noted movement limitations in lower limb joints in SMA patients.<sup>8,24</sup> Johnson et al. described the limitation of extension in 42% out of 20 SMA patients adopting the limitation at the level of minimum 5° as a definition of contracture.<sup>3</sup> With such a definition of a contracture, the limitation of the range of motion was noted in 86% of SMA participants in our observation. These results confirm the need for contracture prevention and stretching in patients with SMA included in the guidelines of care,<sup>10</sup> although the effectiveness of stretching in neurology has not been clearly demonstrated.<sup>25</sup>

The fact that the ranges of HE in SMA individuals are significantly higher in abduction than in the sagittal plane is the most important observation in our study. To date, no one has ever compared the ranges of motion in hip joints in a neutral position and abduction. These differences were particularly visible in the groups of patients with SMA1 and SMA2, while in the SMA3 group mean values of HE in both positions were

similar. Therefore, mainly non-walking individuals were at risk of the occurrence of differences between the ranges of HE in a neutral position and in abduction. It may result from the habit of abduction and external rotation in lower limbs, increased tension in the iliotibial band, weaker hip adductor muscles or an improper build of hip joints. Further research is necessary to explain this phenomenon.

The obtained results indicate the need to apply support standing with abduction in the hip joints in SMA individuals. Previous guidelines for pediatric supported standing programs have recommended standing with hip abduction;<sup>20,21</sup> however, mainly children with cerebral palsy were examined. Mechanics of hip joints in SMA patients is different than in other diseases, especially in cerebral palsy, due to significant differences in muscle tone, lower limb position and motor capacity.

The study revealed that higher limitations of HE occurred in SMA1 and SMA2 participants than in individuals with SMA3. Differences may result from the functional state of patients with particular types of SMA but they may also be related to the duration and frequency of maintaining a standing position. Several authors have described serious limitations in patients with SMA2 and considerably lower limitations in walking patients.<sup>8,11,24,25</sup> They also noted the correlation between the limitations of the ranges of motion in limb joints and the level of general fitness which depends on the type of SMA.

Our study confirmed the positive impact of supported standing on the ranges of motion in the hip joints. The researchers pointed to the fact that contractures constituted a problem for SMA patients who lost the ability to walk.<sup>11,26,27</sup> A positive influence of supported standing was noted in boys with Duchenne muscular dystrophy. It was revealed that supported standing in long knee-ankle-foot orthoses lengthens the time of unsupported walking in boys.<sup>28,29</sup> McDonald et al. noted that boys with Duchenne muscular dystrophy who underwent supported standing were at a lower risk of scoliosis and increasing contractures.<sup>16</sup> A mean duration of supported standing in our study was close to the recommended one.<sup>10</sup>

The values of HE were related to the age of the study participants. The individuals aged more than 6 demonstrated significantly lower ranges of motion than younger subjects. Other authors also noted an increase in movement limitations occurring with age.<sup>8,10,24,26</sup> In the past it was noted that a visible movement limitation occurred as early as in the second year of life, and it increased with time and often needed surgical treatment.<sup>8</sup>

Our observations revealed an interdependence between the difference in the ranges of HE and scoliosis. Scoliosis occurred in 53% of the study participants, which constituted a lower frequency of occurrence compared to other findings.<sup>30,31</sup> Lower occurrence must result from the fact that the group included as many as 22 children aged 2–3 at which age scoliosis is diagnosed less frequently. Bigger differences between the ranges of extension in hip joints were noted in individuals with scoliosis. It indicates a biomechanical relationship between the position of the spine, pelvis and hip joints. Several researchers have been drawing attention to the correlation

between the shape and function of hip joints and scoliosis. While examining mainly patients with SMA2, Granata et al. described the linear correlation between the shift of a femoral bone head and scoliosis.<sup>32</sup> Canavese and Sussman noted that in the majority of individuals with a neuromuscular disease, subluxation or dislocation in the hip joint occurred on the side where pelvis is elevated.<sup>7</sup>

## 6. CONCLUSIONS

- (1) The range of extension in hip joints in SMA patients depends on the hip position. Abduction increases the range of extension.
- (2) Supported standing with hip joint abduction should be used in SMA patients, depending on the value of extension in hip joints in the sagittal plane and abducted position, and on structural changes in the hip joint.
- (3) The results have a practical aspect due to new forms of pharmacological treatment that improve the patient's motor status.

## Conflict of interest

The authors declare that there is no conflict of interest.

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## Ethics

The study was accepted by the Senate Research Ethics Committee at Józef Piłsudski University of Physical Education (SKE 01-03/2016).

The parents of the children and the adults participating in the study were informed about the aims and signed the consent to participate in the study.

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