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Peeters, Laura H C; Janssen, Mariska M H P; Kingma, Idsart; van Dieën, Jaap H; de Groot, Imelda J M

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Patients With Spinal Muscular Atrophy Use High Percentages of Trunk Muscle Capacity to Perform Seated Tasks

Laura H. C. Peeters, MSc, Mariska M. H. P. Janssen, PhD, Idsart Kingma, PhD,
Jaap H. van Dieën, PhD, and Imelda J. M. de Groot, PhD

Objective: The aim of the study was to investigate trunk function during seated upper limb tasks in patients with spinal muscular atrophy types 2 and 3.

Design: Seventeen persons with spinal muscular atrophy and 15 healthy controls performed several tasks when sitting unsupported, such as reaching (and placing) forward and sideward. Joint torque and muscle activity were measured during maximum voluntary isometric contractions. Three-dimensional kinematics and normalized muscle activity were analyzed when performing tasks.

Results: Trunk joint torques were significantly decreased, approximately 45%, in patients with spinal muscular atrophy compared with healthy controls. Active range of trunk motion was also significantly decreased in all directions. When performing tasks, the average back muscle activity was 27% and 56% of maximum voluntary isometric contractions for healthy controls and spinal muscular atrophy and for abdominal muscles 10% and 44% of maximum voluntary isometric contractions, respectively. Trunk range of motion did not differ when performing daily tasks.

Conclusions: The trunk of patients with spinal muscular atrophy is weaker compared with healthy controls, reflected by reduced trunk torques and decreased active range of motion. In addition, patients with spinal muscular atrophy use high percentages of their trunk muscle capacity to perform tasks. Clinicians should take this into account for intervention development, because using high percentages of the maximum muscle capacity results in fatigue and muscle overloading.

Key Words: Muscular Atrophy, Spinal Torso, Activities of Daily Living, Electromyography

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From the Department of Rehabilitation, Donders Centre for Neuroscience, Radboud University Medical Center, Nijmegen, the Netherlands (LHCP, MMHPJ, IJMdG); and Department of Human Movement Sciences, Faculty of Behavioral and Movement Sciences, Vrije Universiteit Amsterdam, Amsterdam Movement Sciences, Amsterdam, the Netherlands (IK, JHvD).

All correspondence should be addressed to: Laura H.C. Peeters, MSc, Department of Rehabilitation, Radboud University Medical Center, PO Box 9101, 6500 HB Nijmegen, the Netherlands.

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This article about trunk movement of the healthy controls was published. This group is also used to compare the SMA patients. However, the focus of this article was on the movement of individual trunk segments, whether this article describes, and compares the trunk as one segment. Therefore, figures, or data, do not overlap.

Laura H. C. Peeters is in training.

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Spinal muscular atrophy (SMA) is characterized by progressive degeneration of motor neurons in the spinal cord, leading to muscle weakness and atrophy.¹ As a result, patients experience limitations in performing daily activities independently.^{2,3} Patients are categorized based on maximum acquired milestones and disease onset, but clinically, it is more a gradual scale of functional abilities.^{4,5} The natural course of children with SMA is characterized not only by weakness of upper and lower limb muscles but also by (severe) weakness in the trunk leading to scoliosis at young age.⁴ However, the natural course is now changing because of effective treatment with Spinraza.⁶

When performing seated activities, the trunk plays an indispensable role as it interacts with upper limb (UL) movement as part of the kinematic chain and it provides a stable base for UL movements.^{7–10} Only a few studies describe trunk function in patients with SMA. Trunk muscle force and axial function seems to be less for patients with SMA type 2 compared with type 3.^{4,11} However, the literature contradicts whether axial function decreases with age. Vuillerot et al.⁵ found only a decline in axial motor function for SMA type 2 patients based on the motor function measure dimension 2, whereas Wadman et al.¹¹ found a decline in motor function for all SMA types based on the Hammersmith Functional Motor Score. Both measures are not solely based on axial function (ie, upper or lower limb function also influences the score), which might explain differences in findings. It is remarkable that so little research has been done concerning trunk function, although scoliosis secondary to muscle weakness is a major problem in childhood for patients with SMA.⁴

Therefore, the aim of this study was to investigate trunk function and its relation with UL movements when performing seated UL tasks in patients with SMA types 2 and 3. We hypothesized that maximum trunk torques and maximum active range of movement are reduced in patients with SMA (types 2 and 3) compared with healthy control (HC), whereas trunk movements and muscle activity levels when performing daily tasks are increased to compensate for reduced UL function and trunk muscle strength.

METHODS

Participants

Seventeen people with SMA and 15 HCs participated in this study. Participants were included if they were older than 6 yrs, able to bring their hand to the mouth, and could sit independently (without back or arm rests) for at least 5 mins. Patients also needed to have a genetically confirmed diagnosis of SMA. Participants were excluded if they had (other) diseases affecting arm, trunk, or head movements.

The 15 HCs were a sample of the HC group described previously.⁹ Because our participants with SMA were mainly adults, we selected only HCs older than 12 yrs to eliminate the maturation effect (eg, coordination between trunk and arm movements changes in children up to the age of 10 yrs) as previously described.^{9,12} For the same reason, the 6-yr-old participant with SMA (SMA_6y) will be described and compared separately with 3 HCs of 6 yrs (HC_6y) as a case study.

Participants with SMA were recruited through advertisements by patient organizations (Spierziekten Nederland and Prinses Beatrix Spierfonds) and through the Radboudumc outpatient clinic Nijmegen. Healthy controls were recruited from local primary schools, high schools, and university. Written informed consent was given by all participants before participation. The study was approved by the medical ethics committee Arnhem-Nijmegen (NL58988.091.16) and all data were handled according to the guidelines of good clinical practice. This study conforms to all STROBE guidelines and reports the required information accordingly (see Supplemental Checklist, Supplemental Digital Content 1, <http://links.lww.com/PHM/A834>).

Procedures

We used the same procedure as that used in a previous study with healthy children.⁹ All participants were seated on a height adjustable chair without backrest or armrest. The sitting height was adjusted so that the knees were flexed 90 degrees and both feet were flat on the ground.

First, to determine maximum trunk range of motion (ROM), participants were asked to perform a maximum active flexion movement of their trunk from a seated position, immediately followed by a maximum active extension movement of their trunk (keeping both feet on the ground). The same was done for maximum axial rotation and lateral bending. Thereafter, several reaching (and placing) tasks were performed with a preferred hand at shoulder height: reaching forward, sideways, and contralaterally. Reaching distance and object weight were varied, resulting in the following combinations for forward, lateral, and contralateral reaching: nearby-0 gram ("N-0"), nearby-500 gram ("N-500"), and far-0 gram ("F-0"). Contralateral reaching was not performed at a far distance. Nearby was defined as the distance that could be reached with the arm without moving the trunk (ie, 100% arm length for HCs but could be closer for SMA) and far was defined as 133% of arm length when possible, otherwise as maximum reaching distance. Furthermore, subjects were asked to perform two daily tasks: displace a porcelain plate (circa 600 grams) from left to right on a table with both hands ("Plate") and bring a cup of 200 grams to the mouth ("Drink"). No instructions were given on how to perform the tasks.

Outcome Measures

Data acquisition and analysis were similar as used in a previous study with healthy children and patients with Duchenne muscular dystrophy and will be described briefly.^{9,13}

Participant Characteristics

Patient characteristics were recorded based on self-reports and included age, weight, height, arm preference, age of diagnosis (if applicable), wheelchair confinement, pain in upper

body at time of participation, scoliosis, and spinal fusion surgery. Sitting height was measured, and for patients with SMA, the Vignos Lower Extremity Scale¹⁴ and Brooke Upper Extremity Scale¹⁵ were used for clinical assessment of leg and arm function, respectively.

Three-Dimensional Motion Analysis

An optical motion capture system (Vicon Motion Systems Ltd, Oxford, United Kingdom) was used to record 13 single reflective markers, which were placed on the skin to define positions and orientations of the trunk and pelvis during task performance. Markers on the spinous processes of C7, T6, T12, and L3, a laterally placed marker at level L1/L2, jugular notch, and xiphoid process of the sternum defined three trunk segments (upper thoracic, lower thoracic, and lumbar).⁹ The pelvis markers were placed according to the Vicon Plugin-Gait model with two additional markers on the iliac crest. The markers divided the trunk initially into three segments, because the trunk cannot be seen as rigid segment. However, to make the data more concise, we decided to report the trunk movement as one segment (ie, summation of the three segment angles and pelvis) in this article. Distribution of movement patterns over the individual trunk segments was essentially the same among HCs and patients with SMA without spinal fusion surgery.

All kinematic data were filtered with a bi-directional fourth-order Butterworth low-pass filter (cutoff frequency of 6 Hz). Trunk joint angles are expressed relative to the global coordinate system.

In all three planes of movement, maximum trunk joint angles were determined when performing the active range of trunk motion tasks. For the reaching tasks and daily tasks, the trunk ROM between the start and end of the task (eg, time where wrist velocity exceeded/got less than 5% of its peak velocity) was determined.

Joint Torque and Surface Electromyography

Surface electromyography (sEMG) (Zerowire EMG, Aurion, Italy) was used to measure muscle activity at a sample rate of 1000 samples/sec. Electrodes were placed on the following muscles on both sides of the body: iliocostalis (6 cm from spinous processes of L1), longissimus (3 cm from spinous processes of L3), external oblique (3 cm from axillae midline at height of umbilicus), and medial deltoid (1/3 on the line from acromion to lateral epicondyle of the elbow).^{16,17} The deltoid muscles were included to get an estimate for shoulder muscle effort when performing tasks. Electrodes on the iliocostalis muscle were not placed in two smaller participants with SMA, because of space limitations on the back.

Maximum force was measured using an adjustable static frame myometer with a KAP-E Force Transducer (range = 0.2–2000 N) (Angewandte System Technik, Dresden, Germany). The force signal was sampled at 1000 samples/sec and filtered with a bi-directional fourth-order low-pass filter of 30 Hz. Afterward, the maximum joint torque was calculated by multiplying the measured force with the segment length (ie, moment arm).

Maximal voluntary isometric contractions (MVICs) were performed to determine maximal joint torques and corresponding sEMG amplitudes. Participants' positions for MVIC

measurements were adapted to seated positions, so all participants with SMA could perform the measurements. Two MVIC efforts were performed for 3 secs by the participants for each of the following directions: trunk flexion, trunk extension, lateral bending trunk (left and right), and shoulder abduction (left and right) (Fig. 1). When the maximum force of the MVIC task varied more than 10% between the two trials, an additional trial was recorded. Because patients with SMA are easily fatigued, it was not feasible to perform many MVIC trials.

A fourth-order Butterworth filter (20–450 Hz) was used to filter the sEMG signals, followed by rectification and low-pass filtering (3 Hz) of the signals to obtain the linear envelopes. The maximum sEMG amplitude for each trunk muscle was taken as the highest amplitude from the four MVIC tasks of the trunk and maximum deltoid sEMG amplitude as the highest amplitude from the shoulder abduction task. Normalized sEMG amplitudes (maximum sEMG amplitude during task divided by

the maximum MVIC amplitude for that muscle) were used to describe the percentage of muscle capacity used during maximum active ROM and daily tasks. Subsequently, average normalized muscle activity of the back muscles (ie, longissimus and iliocostalis both sides) and average normalized activity of the abdominal muscles (ie, external oblique both sides) were calculated. If more than two values were missing, because of inability of the participant to perform the task or because of technical errors, such as missing signals as result of loose electrodes, the average normalized muscle activity was defined as missing value.

All analyses were performed using custom scripts in Matlab R2014b (MathWorks, Natick).

Statistics

Median values and interquartile ranges are used to describe the data because the data were not normally distributed. Wilcoxon rank-sum tests were used to assess differences

FIGURE 1. Participant's positioning for the maximum voluntary isometric contraction tasks with the static frame myometer (indicated by arrow). Top row: trunk flexion and extension; bottom row: lateral bending and shoulder abduction.

TABLE 1. Participant characteristics

	HCs			SMA Patients		
	<i>n</i>	Median	IQR	<i>n</i>	Median	IQR
Age, yr	15	18.1	(14.4–20.4)	16	43.5	(25.5–57.2)
Sex, male/female	7/8			11/5		
Weight, kg	15	60.0	(51.1–66.5)	16	74.5	(56.6–88.1)
Height, cm	14	170.5	(166.0–174.0)	15	176.0	(167.0–178.8)
Sitting height, cm	15	64.5	(62.1–67.5)	14	63.0	(52.0–69.0)
Pain at time of participation, <i>n</i>	0			5		
Type of SMA, type 2/type 3				5/11		
Age of diagnosis, yr				16	3.5	(2–16)
Wheelchair confinement, <i>n</i>				14		
Scoliosis, <i>n</i>	0			9		
Spinal fusion surgery, <i>n</i>	0			6		
Vignos lower extremity scale	1	2	9			
<i>n</i>	1	1	14			
Brooke upper extremity scale	1	2	3	4	5	6
<i>n</i>	3	3	8	0	1	1

IQR, interquartile range.

between patients with SMA and HCs. The ROM is depicted in graphs, where the boxes represent 25th, 50th, and 75th percentile, whiskers minimum and maximum nonoutlier values, and dots indicate outliers (more than 1.5 times the interquartile range). All statistical analyses were performed using Matlab R2014b and the statistical significance level was set at $\alpha = 0.05$.

RESULTS

Participant Characteristics

Participant characteristics are described in Table 1. Participants, who reported pain at time of the measurement, had mainly (chronic) shoulder pain, which did not have major impact on their mobility in daily life. None of the participants used medication described as affecting SMA, except for one participant who used Mestinon. The 6-yr-old, type 2 SMA participant is not included in the table. His Vignos scale was 9, Brooke scale 1, and he had no scoliosis.

Three participants with SMA were not able to sit unsupported, perform tasks at the same time, and were excluded from the kinematic and muscle activity analysis. They all had SMA type 2, spinal fusion surgery, and scored 3, 5, and 6 on the

Brooke scale. One of these subjects wore a trunk brace, and others did not.

Joint Torque

Trunk joint torques were significant lesser ($P < 0.01$) for patients with SMA compared with HCs in all directions, with median values slightly less than 50% of HCs (Table 2). Median shoulder torques were less than 25% of HCs in patients with SMA. Spinal muscular atrophy type 2 patients seemed weaker compared with type 3 patients, although the numbers were too small for statistical testing.

Active ROM Tasks

The numbers of participants with missing values in trunk ROM and muscle activity outcomes are shown in Table 3. Maximum active trunk angles were significantly lower ($P < 0.01$) in all directions in patients with SMA compared with HCs (Fig. 2A). Median trunk flexion angle was reduced the most, approximately by 58 degrees, followed by axial rotation (36 degrees), extension (27 degrees), and lateral bending (24 degrees). There was no significant difference between lateral bending and axial rotation to the dominant or nondominant side.

TABLE 2. Maximum trunk joint torque (in Newton meter) in four directions

	HCs			SMA			<i>P</i> HC/SMA
	<i>n</i>	Median	IQR	<i>n</i>	Median	IQR	
Trunk, flexion	15	54.4	(47.8–70.7)	13	23.7	(20.7–34.4)	0.001
Trunk, extension	15	59.6	(46.8–84.3)	13	25.9	(11.2–47.7)	0.001
Trunk, lateral bending D	15	66.4	(52.7–86.9)	13	29.2	(17.7–42.8)	0.005
Trunk, lateral bending ND	15	63.1	(47.5–75.0)	13	31.6	(16.2–42.7)	0.004
Shoulder abduction, D	15	47.9	(35.4–57.0)	16	11.3	(5.1–17.4)	<0.001
Shoulder abduction, ND	15	42.1	(31.0–54.5)	14	9.8	(4.9–13.0)	<0.001

D, toward dominant side; ND, toward nondominant side.

TABLE 3. The number of participants (SMA/HC) who accomplished a task and included data for trunk kinematics, back, and abdominal muscle activity

Tasks	Task Accomplished	Trunk Kinematics	Back Muscle Activity	Abdominal Muscle Activity
Maximum active ROM				
Flexion	13/15	11/15	12/15	13/15
Extension	13/15	11/14	12/15	13/15
Lateral bending	13/15	12/15	12/15	12/11
Axial rotation	13/15	13/15	12/15	11/11
Reaching forward				
N-0	13/15	13/15	12/15	13/15
N-500	7/15	7/15	7/15	7/15
F-0	9/15	8/15	8/15	9/15
Reaching lateral				
N-0	11/15	10/15	10/15	11/15
N-500	7/15	7/15	7/15	7/15
F-0	7/15	7/15	7/15	7/15
Reaching contra-lateral				
N-0	13/15	13/15	12/15	13/15
N-500	7/15	7/15	7/15	7/15
Plate	13/15	13/15	12/15	13/15
Drink	12/15	9/15	11/15	12/15

0, without weight; 500, 500-gram object; F, far; N, near.

Normalized muscle activity levels when flexing and extending were not different between patients with SMA and HCs for the muscles primarily counteracting gravitational moments (eg, back muscles for trunk flexion, abdominal muscles for trunk extension) (Fig. 2B). However, there was a significant increase ($P < 0.01$) in normalized antagonistic activation for patients with SMA, that is, the abdominal muscles for flexion (up to 29% MVIC), back muscles for extension (up to 24% MVIC), and ipsilateral back muscles for lateral bending (up to 39% MVIC). Normalized muscle activity of the ipsilateral back and

abdominal muscles were also significantly greater ($P < 0.05$) for axial rotation (up to 24% MVIC) in patients with SMA.

Performing Daily Tasks

The number of participants who could accomplish a task and the numbers of participants with missing values in trunk ROM and muscle activity outcomes are shown in Table 3. In general, no differences were seen in trunk ROM between patients with SMA and HCs when performing daily tasks (Fig. 3).

FIGURE 2. Rotation angles and muscle activity for active range of trunk rotation tasks. A, Maximum trunk rotation angle when performing a maximum flexion (FLEX), extension (EXT), lateral bending (LAT), or axial rotation (ROT) movement. Lateral bending and axial rotation are mean values toward dominant and nondominant side. B, Maximum muscle activity levels when performing a maximum flexion, extension, lateral bending to dominant side, and axial rotation to dominant side movement. D, dominant side; EO, external oblique; IC, iliocostalis; LO, longissimus; ND, nondominant side. * $P < 0.05$, ** $P < 0.01$.

FIGURE 3. Trunk ROM in patients with SMA and HCs when performing tasks. 0, without weight; 500, 500-gram object; F, far; N, near. * $P < 0.05$, ** $P < 0.01$.

Normalized muscle activity levels for back and abdominal muscles were significantly greater ($P < 0.01$) in patients with SMA compared with HCs for all tasks, except reaching laterally far (Fig. 4). Unsupported static sitting required already three times as much normalized trunk muscle activity for patients with SMA. When performing the daily tasks, the average back muscle activity was 27% of MVIC for HCs and 56% of MVIC for SMA, and the average abdominal muscle activity was respectively 10% of MVIC and 44% of MVIC. In addition, median muscle activity for the deltoid muscle was approximately 100% MVIC and was significantly greater ($P < 0.05$) compared with HCs.

A Case of a 6-yr-Old Participant

In general, little differences were found between SMA_6y and HC_6y. Both joint torque and maximum active range of trunk motion were comparable between the 6-yr-old participants. Trunk ROM of SMA_6y was different from the HC_6y in half of the daily tasks. However, both increased and decreased ROM was seen, and in most tasks, the difference was less than 3 degrees. Variability in normalized muscle activity for HC_6y was too large to reliably compare with SMA_6y.

DISCUSSION

This is the first study describing trunk function in SMA in relation to the performance of UL tasks. Demand on trunk muscles is high when performing such tasks, reflected by increased normalized muscle activity levels as hypothesized, but in contrast with our hypothesis, this occurred without an increased trunk ROM.

Trunk joint torque was decreased in patients with SMA compared with HCs with at least a factor two in median value. In addition, SMA type 2 patients seemed weaker in trunk torque compared with type 3, as was also found previously.⁴ On the other hand, the large interquartile ranges indicate a gradual scale in trunk function, which is in line with the fact that SMA shows a range of functional abilities rather than absolute differences between types of SMA.⁵ More patients are needed to confirm whether there is a difference between types or that it is a graduate scale.

Maximum active trunk ROM was limited in patients with SMA compared with HCs in all directions. To perform the ROM tasks, both groups used a comparable percentage of their maximum muscle capacity for the muscles counteracting gravitational moments in flexion, extension, and lateral bending movements. This indicates that patients with SMA achieve a lower maximum ROM when using similar muscle effort of the counteracting gravitational muscles as HCs. This is not surprising, because the maximum absolute muscle activity is much less for patients with SMA because of loss of motor neurons. A lower maximum absolute muscle capacity results in less force generating capacity, as reflected in the decreased joint torques.

When performing reaching and daily tasks, patients with SMA used a greater percentage of their maximum trunk muscle capacity compared with HC, although trunk movement did not increase. We expected to find increased trunk movement to compensate for reduced arm function, as for example was visible in patients with Duchenne muscular dystrophy.¹³ However, although deltoid muscle activity level was close to 100% of MVIC, trunk ROM did not increase. As a consequence, patients

FIGURE 4. Muscle activity in patients with SMA and HCs when performing tasks. 0, without weight; 500, 500-gram object; F, far; N, near. * $P < 0.05$, ** $P < 0.01$.

will be restricted in their workspace and therefore in performing daily activities. The fact that patients with SMA did not increase their trunk ROM, although normalized shoulder muscle activity was very high, suggests that patients with SMA need more of their trunk muscle capacity to maintain stability to perform the UL movements.¹⁸

To gain more insight in mechanisms underlying the increased normalized muscle activity when performing the reaching and daily tasks, we analyzed the absolute muscle activity. This showed similar absolute muscle activity levels of the back muscles, indicating comparable back muscle activation during task performance in SMA and HCs (in combination with comparable trunk ROM). Noteworthy, this still resulted in increased percentages of normalized muscle activity in patients with SMA because the absolute maximum muscle activity was decreased. On the other hand, the absolute abdominal muscle activity was significantly increased in patients with SMA, which could indicate co-contraction of the abdominal muscles during task performance and would support the hypothesis above. The co-contraction can be caused by recruitment of more motor units needed to generate enough muscle force to maintain trunk stability and/or recruitment of larger motor units because of re-innervation in SMA.¹⁹

Using increased percentages of the maximum muscle capacity and co-contraction causes earlier development of fatigue and increased risk of muscle overloading.^{18,20} Because scoliosis is related to muscle weakness and fatigue, clinicians should pay high attention to trunk function in children with SMA.²¹ However also in general for functional assessment and development of interventions, there should be more awareness for the great loads on trunk muscles required to perform simple manual tasks. Interventions to reduce muscle fatigue during the day can be applied, such as proper seating, use of trunk

supportive devices, or physical muscle strength training to reduce fatigability.^{22,23} Rigid trunk orthoses are not recommended, because these restrict important trunk movements that are necessary to perform daily tasks. In addition, being able to move could also prevent the muscles from degenerating faster because of disuse.^{3,24} New supportive devices that allow movement and reduce load on the trunk are needed.

For the first time in patients with SMA, a quantitative insight in trunk function was obtained. The results were consistent with clinical experience on trunk function and can therefore support clinical decision-making. Furthermore, the method used in this study gives opportunities to evaluate interventions in a quantitative manner in the future. Treatment with, for example, Spinraza is currently evaluated with the use of the Hammersmith Functional Motor Scale, but this does not discriminate between different body segments and does not give insight in the benefits for performing activities of daily living.^{6,25}

This study has several limitations. First, although we covered a broad range of the clinical spectrum of SMA, it was statistically not possible to compare, for example, SMA type 2 or type 3 patients, or patients with or without spinal fusion surgery because of the small sample size. It would be interesting to investigate in more detail how differences between subtypes affect task performance. Secondly, the control group was not age matched with the patients with SMA. This might have had an effect on the maximum joint torque and maximum active trunk ROM, as muscle strength and joint flexibility decrease with ageing (starting approximately 50 yrs).^{26,27} However, differences found between the HCs and patients with SMA were very high and cannot be solely attributed to age. Furthermore, the reported ROM values during the maximum ROM tasks are active ranges based on unsupported seating, and it should be noted that several participants reported that

they were afraid of falling when moving further. Lastly, the percentages presented for normalized muscle activity are likely an overestimation, because standardized MVIC tasks were performed from a seated position, which likely resulted in lower absolute maximum muscle activity signals. However, this position was chosen, so patients could perform the MVIC tasks and because it corresponded with the position in which the movement tasks were performed.

In conclusion, because of degeneration of motor neurons, patients with SMA need a greater percentage of their maximum muscle capacity to generate the same amount of force as HCs. This study was the first to quantify the effects of this in performance of seated tasks. Maximum trunk joint torque and active trunk ROM were significantly reduced in patients with SMA. Furthermore, increased normalized trunk muscle activity, without increased trunk ROM, was seen when performing daily tasks. Co-contraction of the trunk muscles is very likely present. This indicates that patients with SMA use more of their muscle capacity to maintain trunk stability compared with HCs. Clinicians should take trunk function into account when assessing function and interventions, because using a high percentage of the maximum muscle capacity may result in fatigue and muscle overloading. On the other hand, one must bear in mind that restrictions in trunk movement will likely cause limitations in accomplishing tasks independently and might accelerate muscle decline because of disuse.

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REFERENCES

1. Arnold WD, Kassir D, Kissel JT: Spinal muscular atrophy: diagnosis and management in a new therapeutic era. *Muscle Nerve* 2015;51:157–67
2. Rouault F, Christie-Brown V, Broekgaarden R, et al: Disease impact on general well-being and therapeutic expectations of European Type II and Type III spinal muscular atrophy patients. *Neuromuscul Disord* 2017;27:428–38
3. Bergsma A, Janssen MMHP, Geurts ACH, et al: Different profiles of upper limb function in four types of neuromuscular disorders. *Neuromuscul Disord* 2017;27:1115–22
4. Carter GT, Abresch RT, Fowler WM Jr, et al: Profiles of neuromuscular diseases. Spinal muscular atrophy. *Am J Phys Med Rehabil* 1995;74:S150–9
5. Vuillerot C, Payan C, Iwaz J, et al., MFM Spinal Muscular Atrophy Study Group: Responsiveness of the motor function measure in patients with spinal muscular atrophy. *Arch Phys Med Rehabil* 2013;94:1555–61
6. Mercuri E, Darras BT, Chiriboga CA, et al: Nusinersen versus sham control in later-onset spinal muscular atrophy. *N Engl J Med* 2018;378:625–35
7. Bouisset S, Do MC: Posture, dynamic stability, and voluntary movement. *Neurophysiol Clin* 2008;38:345–62
8. Flatters I, Mushtaq F, Hill LJ, et al: Children's head movements and postural stability as a function of task. *Exp Brain Res* 2014;232:1953–70
9. Peeters LHC, Kingma I, Faber GS, et al: Trunk, head and pelvis interactions in healthy children when performing seated daily arm tasks. *Exp Brain Res* 2018;236:2023–36
10. Robertson JV, Roby-Brami A: The trunk as a part of the kinematic chain for reaching movements in healthy subjects and hemiparetic patients. *Brain Res* 2011;1382:137–46
11. Wadman RI, Wijngaarde CA, Stam M, et al: Muscle strength and motor function throughout life in a cross-sectional cohort of 180 patients with spinal muscular atrophy types 1c–4. *Eur J Neurol* 2018;25:512–8
12. Schneiberg S, Sveistrup H, McFadyen B, et al: The development of coordination for reach-to-grasp movements in children. *Exp Brain Res* 2002;146:142–54
13. Peeters LHC, Kingma I, van Dieën JH, et al: Don't forget the trunk in Duchenne muscular dystrophy patients: more muscle weakness and compensation than expected. *J Neuroeng Rehabil* 2019;16:44
14. Vignos PJ Jr., Archibald KC: Maintenance of ambulation in childhood muscular dystrophy. *J Chronic Dis* 1960;12:273–90
15. Brooke MH, Griggs RC, Mendell JR, et al: Clinical trial in Duchenne dystrophy. I. The design of the protocol. *Muscle Nerve* 1981;4:186–97
16. Hermens HJ, Freriks B, Disselhorst-Klug C, et al: Development of recommendations for SEMG sensors and sensor placement procedures. *J Electromyogr Kinesiol* 2000;10:361–74
17. Willigenburg NW, Kingma I, van Dieën JH: Center of pressure trajectories, trunk kinematics and trunk muscle activation during unstable sitting in low back pain patients. *Gait Posture* 2013;38:625–30
18. Granata KP, Slota GP, Wilson SE: Influence of fatigue in neuromuscular control of spinal stability. *Hum Factors* 2004;46:81–91
19. Querin G, Lenglet T, Debs R, et al: The motor unit number index (MUNIX) profile of patients with adult spinal muscular atrophy. *Clin Neurophysiol* 2018;129:2333–40
20. Nordlund MM, Thorstensson A, Cresswell AG: Central and peripheral contributions to fatigue in relation to level of activation during repeated maximal voluntary isometric plantar flexions. *J Appl Physiol (1985)* 2004;96:218–25
21. Tsirikos AI, Baker ADL: Spinal muscular atrophy: classification, aetiology, and treatment of spinal deformity in children and adolescents. *Curr Orthop* 2006;20:430–45
22. Deering RE, Senefeld JW, Pashibin T, et al: Muscle function and fatigability of trunk flexors in males and females. *Biol Sex Differ* 2017;8:12
23. Mahmood MN, Peeters LHC, Paalman M, et al: Development and evaluation of a passive trunk support system for Duchenne muscular dystrophy patients. *J Neuroeng Rehabil* 2018;15:22
24. Jansen M, van Alfen N, Geurts AC, et al: Assisted bicycle training delays functional deterioration in boys with Duchenne muscular dystrophy: the randomized controlled trial “no use is disuse”. *Neurorehabil Neural Repair* 2013;27:816–27
25. O'Hagen JM, Glanzman AM, McDermott MP, et al: An expanded version of the Hammersmith Functional Motor Scale for SMA II and III patients. *Neuromuscul Disord* 2007;17:693–7
26. Metter EJ, Conwit R, Tobin J, et al: Age-associated loss of power and strength in the upper extremities in women and men. *J Gerontol A Biol Sci Med Sci* 1997;52:B267–76
27. Freemont AJ, Hoyland JA: Morphology, mechanisms and pathology of musculoskeletal ageing. *J Pathol* 2007;211:252–9