Compensatory movements during functional activities in ambulatory children with Duchenne muscular dystrophy

Movimentos compensatórios durante atividades funcionais em crianças deambuladoras com distrofia muscular de Duchenne

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ABSTRACT

Objective: During the transitional phase (ambulatory to non-ambulatory), synergies characterize the evolution of Duchenne muscular dystrophy (DMD). This study was performed to describe and quantify compensatory movements while sitting down on/rising from the floor and climbing up/down steps. **Method:** Eighty videos (5 children × 4 assessments × 4 tasks) were recorded quarterly in the year prior to gait loss. Compensatory movements from the videos were registered based on the Functional Evaluation Scale for DMD. **Results:** The most frequently observed compensatory movements were upper limb support on lower limbs/floor/handrail during all the tasks and lumbar hyperlordosis, trunk support on handrail, equinus foot, increased base of support, non-alternated descent, and pauses while climbing up/down steps. **Conclusion:** Climbing up/down steps showed a higher number of compensatory movements than sitting down on/rising from the floor, which seemed to be lost before climbing up/down steps in ambulatory children with DMD.

Keywords: Duchenne muscular dystrophy, evaluation, scales.

RESUMO

Objetivo: Durante a fase de transição (deambulador – não-deambulador), sinergias musculares caracterizam a evolução da distrofia muscular de Duchenne (DMD). Este estudo visou descrever e quantificar os movimentos compensatórios durante o sentar/levantar do solo, subir/descer degraus. Oitenta vídeos (5 crianças × 4 avaliações × 4 tarefas) foram gravados trimestralmente durante o ano que antecedeu a perda da marcha. **Método:** Os movimentos compensatórios dos vídeos foram registrados utilizando a Escala de Avaliação Funcional para DMD. **Resultados:** Os movimentos compensatórios mais frequentemente observados foram apoio de membros superiores nos membros inferiores/solo/corrimão durante todas as tarefas funcionais e hiperlordose lombar, apoio de tronco no corrimão, pés equinos, aumento da base de suporte, descida não alternada e pausas ao subir/descer degraus. Subir/descer degraus apresentou um número maior de movimentos compensatórios do que sentar/levantar do solo. **Conclusão:** Sentar/levantar do solo foram habilidades perdidas antes de subir/descer degraus em crianças com DMD.

Palavras-chave: distrofia muscular de Duchenne, avaliação, escalas.

Duchenne muscular dystrophy (DMD) is a recessive X-linked form of muscular dystrophy in children, affecting 1 in every 3,500 male births. The disorder is caused by a mutation in the dystrophin gene located on the X chromosome, which causes total or partial absence of this protein on the cell membrane, resulting in irreversible and progressive loss of functional abilities[1,2].

Symptoms usually appear in early childhood, when the child is between 3 and 7 years old. Parents report frequent falls and difficulty running, rising from the floor, and climbing steps. The physical examination shows bilateral symmetrical weakness of proximal muscles, which occurs progressively starting from the lower limbs. Gait loss usually occurs when the child is between 9 and 12 years old[2-5].

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Mercuri *et al.* carried out prospective and retrospective studies to establish the importance of functional scores in detecting alterations, such as gait loss, in DMD[6]. Previous research assessed the reliability and validity of two domains (sitting down on the floor/rising from the floor and climbing up/down steps) of the Functional Evaluation Scale for ambulatory patients with DMD (FES-DMD)[7,8]. These studies provided a scale for the systematized observation of movements of children with DMD. This scale allows the use of videos, which permit analysis of the separate phases of the activities, avoid musculoskeletal overload, and generate a permanent databank. The scale scores the adoption and maintenance of postures (sitting, kneeling, standing, etc.) and the use of compensatory movements.

The qualitative analysis of compensatory movements during functional tasks may allow the recognition of important changes in synergies. This assessment may predict the loss of the ability to perform activities of daily living. Not being able to rise from the floor or climb up/down steps are frequently chief complaints described during physical therapy sessions with ambulatory children with DMD and their caregivers. We hypothesized that these tasks would be lost in the 12-month period preceding the gait loss. We also believed that these tasks would not be lost simultaneously. The description of the sequence of functional loss of these four tasks might clarify the progression of DMD. Therefore, knowing which of these tasks would be lost first might help professionals provide more specific assistance and guidance, with more tailored kinesiotherapy and orientations[9,10].

This study aimed to describe and quantify compensatory movements during sitting down on and rising from the floor and climbing up/down four steps to determine the sequence of functional loss in DMD.

METHOD

This research was approved by the Ethics Committee of University of São Paulo (USP) (project 285/10). It was an observational, descriptive, and retrospective 1-year study.

We observed 80 video assessments: 20 videos of each of the 4 tasks (sitting down on the floor, rising from the floor, climbing up steps, and climbing down steps) recorded at four time points (12, 9, 6, and 3 months) prior to the gait loss. Five children performed the functional tasks (4 tasks × 4 moments × 5 children=80 videos). Compensatory movements were observed based on the FES-DMD. The inclusion criteria were (1) having a record of performing (or at least trying to perform) the tasks: rising from the floor/sitting down on the floor and climbing up/down steps on four quarterly periodical evaluations during the year prior to the gait loss, (2) having Vignos scores available, and (3) having a Vignos score between 1 and 3 at 12 months prior to gait loss.

Two physiotherapists, with a minimum of 2 years' experience with patients with neuromuscular disorders, observed the motor strategies necessary to perform the tasks. They quantified compensatory movements based on the FES-DMD. Videos with technical problems, such as poor illumination or non-visualization of body segments, were excluded.

A descriptive analysis with histograms showing frequency distributions was used to describe compensatory movements. Frequency indexes and quotients were calculated based on the total number of evaluations prior to the gait loss.

The Vignos Scale[11] and two domains of FES-DMD were used: sitting down on/rising from the floor[7], climbing up/down steps and climbing down steps[8]. The absolute number of compensatory movements was also recorded.

The FES-DMD evaluation of sitting on the floor is divided into three phases: (1) trunk flexion, (2) knee and ankle flexion, and (3) sitting position. The activity of rising from the floor is composed of five phases: (1) supine to sitting or side-lying, (2) transition to sitting, (3) transition to kneeling, (4) quadrupedalism, and (5) quadrupedalism to standing[7]. The evaluation of climbing up steps and climbing down steps consists of four phases for both: (1) preparation, (2) propulsion, (3) swing, and (4) stance[8]. In FES-DMD, higher scores indicate a worse performance.

Frequency indexes of each task were created, considering all the evaluations performed by the 5 children ([5 children × 4 evaluations] – evaluations in which one or more patients could not perform the task). Therefore, the frequency index was the total number of times the compensation was observed, considering the five children and the four assessments. As some children were unable to perform all the four tasks up to the end of the study period, these incomplete videos were considered as "not being able to perform the task." Consequently, the total number of videos showing successful attempts for each task was different from 20. In 15 video records, children succeeded sitting down on the floor (in 5 video records, children were not able to perform the task); in 16 videos children performed rising from the floor (in 4 videos, the task was not finished); in 17 videos, children performed climbing up steps; and in 18 videos, they were able to climb down steps.

A frequency quotient was also created, dividing the frequency index by the total number of evaluations performed (the assessments in which the patients were not able to perform the tasks were not considered in this quotient). The frequency quotient was the frequency index divided by the total number of available assessments. The most frequent findings were highlighted to identify possible predictors of gait loss.

Description of outcomes

The Vignos classification varied from 2 to 5. On the first evaluation, children scored 2 or 3, but on the last evaluation three children scored 3 and two children scored 5. The scores

on FES-DMD on "sitting down on the floor" varied from 5 to 10, those for "rising from the floor" varied from 6 to 14, "climbing up steps" varied from 17 to 26, and "climbing down steps" varied from 15 to 23.

The compensatory movements during the activities sitting down on the floor and rising from the floor seem to have a limit. The maximum number of compensatory movements observed per child was 5 for sitting and 6 for rising from the floor. The activities climbing up and down steps did not show this clear limit, and the number of compensatory movements varied from 17 to 27 and from 17 to 25, respectively (Figures 1 and 2).

The compensatory movements are listed in Tables 1 and 2. The most frequent were: "sitting down on the floor," upper limbs support on lower limbs and/or on the floor and transition to side-sitting before sitting; "rising from the floor," transition to side-lying before sitting and stabilizing the trunk with the support of both upper limbs and/or external support; and for "climbing up and climbing down steps," upper limbs support on lower limbs and/or on the handrail, trunk support on handrail, lumbar hyperlordosis, equinus swing foot, increased base of support, non-alternated descent, and pauses after each step (Tables 1 and 2).

DISCUSSION

Motor control is influenced by (1) individual aspects, such as structural characteristics (body mass index, muscle trophism), perception (previous experiences, awareness of body
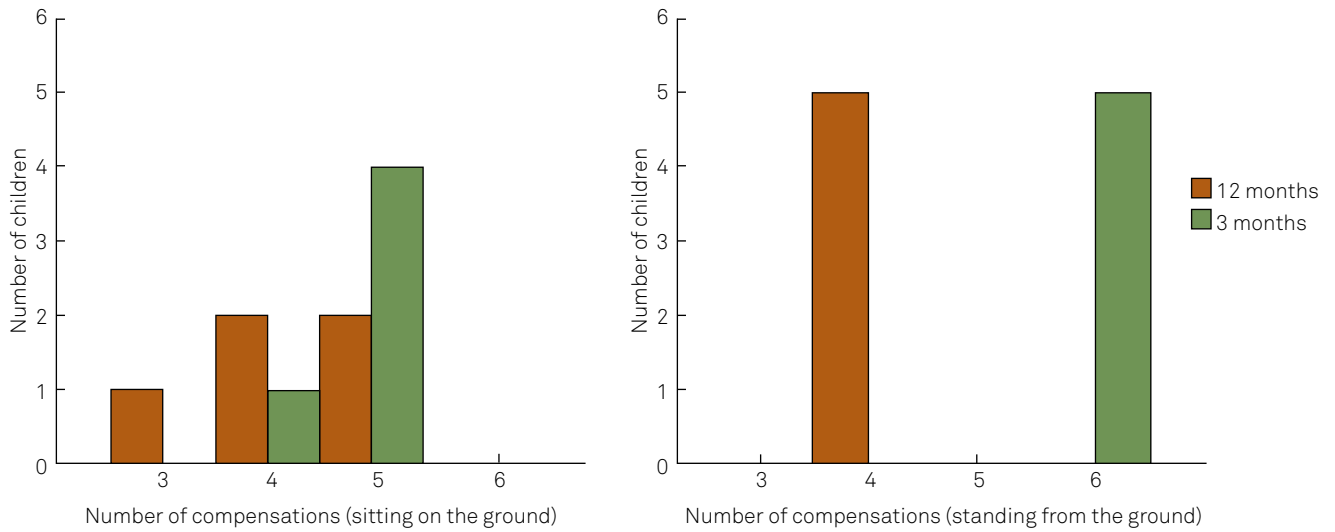

Figure 1. Number of children performing compensatory movements on the tasks "sitting down on the floor" and "rising from the floor" on evaluations 12 months and 3 months prior to gait loss.
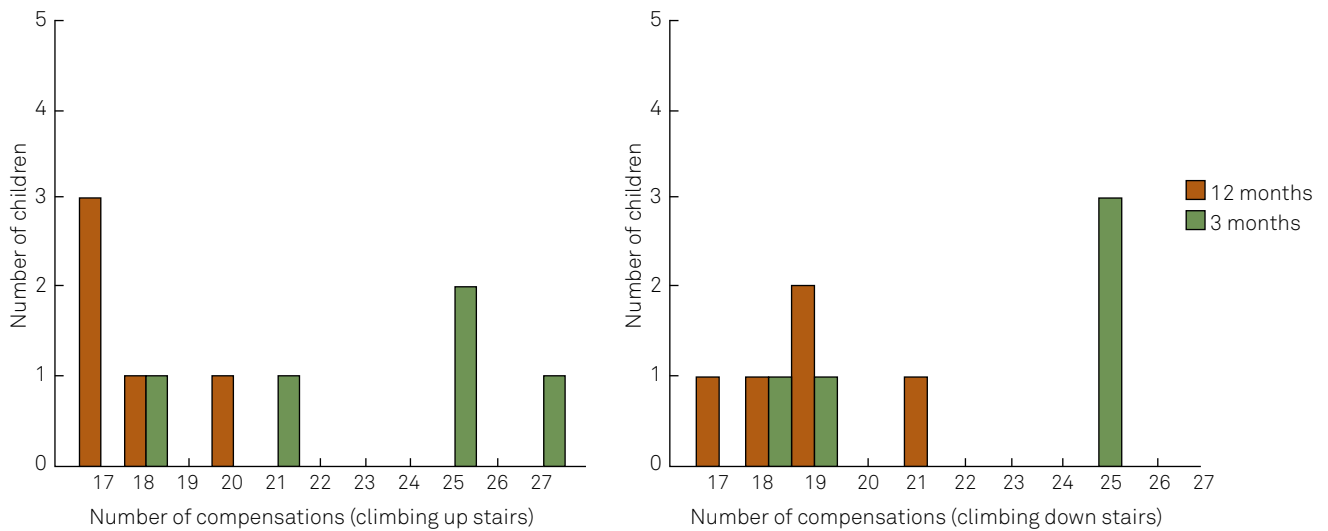

Figure 2. Number of children performing compensatory movements on the tasks "climbing up steps" and "climbing down steps" on evaluations 12 months and 3 months prior to gait loss.

Table 1. Compensatory movements observed on "sitting on the floor" and "standing from the floor". For "sitting on the floor," a total of 16 assessments were considered, and for "standing from the floor," 15 were noted because some children could not perform the tasks on the last assessment (3 months prior to gait loss). The frequency index is the number of times the compensation was observed in the five children during the four assessments. The frequency quotient is the frequency index divided by the total number of assessments.

Sitting on the floor	Motor compensation	Frequency index	Frequency quotient
Phase			
Trunk flexion	Upper limb support on the floor	13	0.81
	Upper limb support on lower limbs and on the floor	3	0.19
Knee/ankle flexion	Upper limb support on lower limbs or on the floor	16	1.00
	Upper limb stabilization	8	0.50
Sitting position	Upper limb stabilization and support	7	0.44
	Side-sitting	13	0.81
	Failure on eccentric control on descent	11	0.69
Standing from the floor			
Supine to sitting or side-lying	Upper limb support	3	0.20
	Supine to side-lying	12	0.80
Transition to sitting	Upper limb support/stabilization	2	0.13
	Both upper limbs support/stabilization	13	0.87
Transition to kneeling	Sitting to quadrupedalism	8	0.53
	Kneeling with external support	7	0.47
Quadrupedalism	Maintenance with body support	8	0.53
	Upper limb support on knees	1	0.07
Quadrupedalism to standing	Upper limb support on knees and thighs	3	0.20
	Upper limb support on legs, knees, and thighs	1	0.07
	External support needed	10	0.67

position in space and ability to integrate sensory information), motor impairment (weakness, joint instability, range of motion, motor coordination), cognition (attention, planning, executive function, motivation, emotional distress, and fear of falling); (2) environmental aspects (furniture, presence of the caregiver); and (3) characteristics of the task (complexity, timing, degrees of freedom, familiarity)[12-14]. To deal with individual, environmental, and task constraints, compensatory motor strategies are usually helpful[12].

In the present study, we observed patients with muscle weakness due to DMD performing tasks considered difficult because they demanded eccentric and antigravity control. Disease progression resulted in the emergence of multiple and varied synergies to compensate muscle weakness and deal with the demands of the tasks.

We verified that sitting down on and rising from the floor seemed to have a limit of possible compensatory movements based on the FES-DMD. On the evaluation performed 3 months prior to gait loss, sitting on the floor showed five possible compensatory movements and rising from the floor was associated with six. However, climbing up and down steps showed a more varied motor repertoire, with no clear limit to possible compensatory movements.

The analysis of sitting down on the floor evidenced the need for the support of upper limbs on the floor during the trunk flexion phase on all evaluations. This result differs from the study of Escorcio et al., which observed that healthy boys

(n=40, mean age: 12) performed this task with trunk anteriorization, hip and knee flexion, and support of the hands behind the pelvis[15].

The analysis of rising from the floor showed the frequent need for external support during the transfer from quadrupedalism to standing. This does not coincide with the normal and frequent movements seen in healthy young adults: the support of the hands on the floor behind the pelvis and a symmetric squatting to assume standing[12,15]. This difference in performance can be explained by many factors, such as difficulty in contracting the hip and trunk extensors and difficulty to transfer to half-kneeling due to hip flexor weakness, and difficulty in standing up when triple extension is required in a closed kinetic chain due to hip, knee, and ankle extensor weakness. The compensatory movement observed in boys with DMD during this task, known as Gowers' sign, is a typical way of dealing with the weakness of the antigravity muscles of the lower limbs.

Climbing up and down steps are complex activities that demand joint stability and correct timing and the selection of muscle synergies. DMD compromises lower limb strength, mainly and firstly hip extensors, resulting in an anterior pelvic tilt and lumbar hyperlordosis. It is followed by quadriceps and ankle dorsal flexors weakness[9]. These impairments result in poorer performances on both tasks.

During locomotion on steps, the movements of the center of gravity in the sagittal and frontal planes are significant;

Table 2. Compensatory movements on "climbing up and down stairs." A total of 17 and 18 assessments were considered, respectively (some children could not perform the tasks on the last assessment 3 months prior to gait loss. The frequency index is the number of times the compensation was observed, considering the five children and the four assessments. The frequency quotient is the frequency index divided by the total number of assessments.

	Motor compensation	Climbing up stairs		Climbing down stairs	
		Frequency index	Frequency quotient	Frequency index	Frequency quotient
Phase	One hand support	1	0.06	2	0.11
	Both hands support	15	0.88	16	0.89
	Forearm support	1	0.06	2	0.11
	Trunk/head rotation	1	0.06	11	0.61
Preparation	Trunk/head lateral bend	1	0.06	3	0.17
	Lumbar hyperlordosis	16	0.94	18	1.00
	Swing knee flexion	1	0.06	1	0.06
	Swing hip internal rotation	3	0.18	1	0.06
	Increased base of support	12	0.71	15	0.83
	Equinus swing foot	16	0.94	18	1.00
	Both hands support	13	0.76	17	0.94
	Other hand on stance knee	1	0.06	1	0.06
Propulsion	Forearm propulsion/support	4	0.24	2	0.11
	Shoulder elevation	1	0.06	4	0.22
	Head rotation/anteriorization	7	0.41	1	0.06
	Trunk flexion	11	0.65	14	0.78
	Trunk extension/hyperlordosis	17	1.00	13	0.72
	Trunk lateral bend/rotation	5	0.29	13	0.72
	Swing pelvis elevation	12	0.71	5	0.28
	Swing hip extension	0	0.00	18	1.00
	Swing hip abduction	9	0.53	1	0.06
	Swing hip internal rotation	12	0.71	12	0.67
Swing	Swing foot extension	11	0.65	18	1.00
	Stance knee flexion	0	0.00	16	0.89
	Stance on forefoot	11	0.65	1	0.06
	Swing foot repeatedly touching the step	6	0.35	2	0.11
	Trunk support	17	1.00	9	0.50
	Stance knee flexion	7	0.41	0	0.00
	Stance knee extension	0	0.00	13	0.72
	Stance foot instability	6	0.35	1	0.06
Stance	Increased base of support	17	1.00	17	0.94
	Pauses after each step	17	1.00	18	1.00
	Non-alternated descent	17	1.00	14	0.78

the base of support is mobile and varies on each step. Although this task requires a higher degree of attention, vision, and coordination, it also involves a higher number of joint degrees of freedom compared to sitting down on and rising from the floor[8,13]. Therefore, it may show a higher variability in performance, which may explain the high number of possible compensatory movements observed in the present study. Individuals can find different ways of dealing with weak muscles. Weakness may become a constraint inducing the selection of new motor strategies. This might explain the maintenance of climbing up/down steps for a longer time compared to sitting down on/rising from the floor[12].

According to McFadyen and Winter[16], hip position directly interferes with trunk position when climbing up and down steps. In the present study, the children had to use hip compensatory movements (lateral elevation, internal rotation, abduction) and also trunk support on the handrail to climb up steps. Sivaraman et al.[17] reported that ankle dorsal and plantar flexion are the most important movements when climbing down steps. This makes this task particularly difficult for children with DMD, who usually have equinus gait and/or reduced range of motion and muscle strength. Antigravity muscles perform eccentric contractions during this task, increasing the risk of fiber damage. Therefore, the use of upper limbs and trunk support may minimize the difficulty of eccentric control and avoid a faster progression of motor impairments[18].

Considering the sequence of functional impairment, one child became unable to sit down on the floor 9 months prior to gait loss, whereas another lost this function 6 months prior to

gait loss. Also, two other children were not able to rise from the floor 3 months before gait loss. Finally, the two children who were unable to sit down on the floor at 9 and 6 months could not climb steps at 3 and 6 months, respectively. Also, they were unable to climb down steps at 3 months prior to the gait loss. Therefore, sitting down on the floor seems to be the first functional task to be lost prior to gait loss, followed by rising from the floor, climbing up steps, and climbing down steps.

It is interesting to observe that one child could not sit down on the floor before he could not rise from the floor, while the other four children first became unable to rise from the floor. Although rising from the floor requires a higher muscular effort, the lack of eccentric control of the antigravity muscles may have resulted in a different progression. During the video assessment of rising from the floor, this child was assisted by the physiotherapist.

It is important to emphasize that most children with DMD could not perform the tasks (sitting down on/rising from the floor, followed by climbing up/down steps) prior to gait loss, showing that the evaluation of these tasks can provide additional clinical information, particularly in the end of the ambulatory period. This will facilitate professional assistance during the transitional phase (from ambulatory to non-ambulatory).

In a study of functional independence in 53 children with DMD, all of them first became unable to rise from a chair (mean age: 9.4 years); after that, 42 of them could not rise from the floor (10.2 years); and finally, all of them could not climb up and down steps (10.4 years) before 44 of them became non-ambulatory (11.4 years). These data agree with our hypothesis of the existence of a sequence of functional impairments prior to gait loss[5]. The present study places the interruption of the ability to sit on/rise from the floor before climbing up/down steps. The ability of sitting on/rising from a chair remains longer than the ability to sit on/rise from the floor. Taking the findings by Brooke et al.[5] and the results described here, the probable sequence of functional loss is the interruption of sitting on/rising from the floor, sitting on/rising from a chair, and finally, climbing up/down steps. However, future studies should simultaneously analyze all of these activities in a longitudinal study with children with DMD.

Studies on scales for patients with neuromuscular diseases emphasize muscle strength, but postulate that there is no direct relationship between strength and functional performance[10,17,19,20]. However, Doglio et al. reported that some motor strategies may be adopted to compensate for muscle weakness and postpone wheelchair dependency[21].

Functional scales indicate disease progression and can help guide therapeutic decisions, which are also based on the general aspects mentioned previously[13,22-24]. The Motor Function Measure (D1-MFM) can detect changes in 3-month intervals: dimension 1 (standing, transfers), when scored from 40% to 70%, can predict the loss of ambulation[22]. Nevertheless, this percentage of variation is wide and makes the prediction of wheelchair dependency difficult.

The Medical Research Council also reports muscle strength lower than 3 in hip extensors and lower than 4 in ankle flexors as a good predictor of gait loss. The inability to climb steps and an increase in the frequency of falls is also a good predictor[5].

Three children did not show any variation on the Vignos scale during the 1-year period of the present study. This reinforces the classification characteristic of this scale and the need for other additional clinical scales, such as the FES-DMD, to guide functional evaluation and therapeutic approaches[16,18,21-23]. The evaluations of sitting down on/rising from the floor, climbing up/down steps with FES-DMD, and the quantification of compensatory movements can help to identify the best moment to prepare the child and their family to present the wheelchair as a safer option for locomotion.

This study has several limitations: (1) a small sample, which can be justified by the difficulty of following children with DMD with periodic evaluations prior to gait loss; and (2) the inability to describe other clinical measures/information, such as body mass index, medication, muscle strength index, and range of motion, because the study used videos from a databank.

In conclusion, children with DMD seem to reach a clearer limit of possible compensatory movements while sitting down on and rising from the floor compared to climbing up and down steps in the year prior to the gait loss. When climbing up and down steps, children show a higher variability of movements, with a less evident limit of possible compensatory movements preceding gait loss. In our sample, most children failed to perform these four tasks 3 months prior to the gait loss. Also, sitting down and rising from the floor seems to be lost before climbing up and down steps in ambulatory children with DMD.

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