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Efficacy of the Head Up collar in facilitating functional head movements in patients with Amyotrophic Lateral Sclerosis

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ABSTRACT

Background: The Head Up collar is a cervical orthosis designed to be adaptable to a patient's needs using adjustable removable supports. The aim of this study was to characterise the ability of this orthosis to provide head support and facilitate the control of head movements in people living with Amyotrophic Lateral Sclerosis.
Methods: Thirteen patients (6 females, age range: 45–74 years old, Amyotrophic Lateral Sclerosis Functional Rating Scale range: 13–44) with neck muscle weakness due to Amyotrophic Lateral Sclerosis were enrolled in the study. An additional inclusion criterion was the presence of enough residual muscle strength to enable the performance of the test procedure. Participants were asked to perform a series of head movements with and without wearing the collar. Two parameters (mean angular velocity and ratio of movement coupling) were extracted from recorded angular velocities, to quantify changes in the execution of the movement between the two conditions.
Findings: Participants exhibited different levels of impairment in performing different movements. When wearing the collar self-selected movement velocity was preserved and significant improvement in the control of lateral flexion movement was observed (median ratio of movement coupling value reduced from 1.1 to 0.84, $P = 0.013$). A lower ratio of movement coupling was also observed in 4 out of 7 individuals that were fitted with anterior supports.
Interpretation: The heterogeneity observed in the level of impairment and residual function highlights the need for personalized interventions. The Head Up was effective in enabling more controlled movements and maintaining the natural velocity of head movement.

1. Introduction

Amyotrophic Lateral Sclerosis (ALS)/motor neuron disease (MND) is a neurodegenerative disease that affects upper and lower motor neurons leading to progressive muscular atrophy and eventually death (Hobson and McDermott, 2016). As with other neuromuscular pathologies (Gourie-Devi et al., 2003), presentations of ALS may include neck muscle weakness and head drop (Martin et al., 2011). Head drop exacerbates problems with swallowing, communicating and breathing, causing significant difficulties in performing activities of daily living and is negatively associated with survival time (Nakamura et al., 2013).

Using a quantitative biomechanical approach, neck movements in ALS patients are characterized by a lower velocity, are less smooth and present an increased amount of so called “coupled movements”, compared to age-matched healthy subjects (Pancani et al., 2017). The latter are undesired out of plane movements that are associated with the intended primary movement. Due to the morphology of the cervical spine, even in healthy individuals, when a gross rotation is performed, a

Abbreviations: ALS, Amyotrophic Lateral Sclerosis; plwALS, Patients living with Amyotrophic Lateral Sclerosis; ALSFRS-R, ALS Functional Rating Scale - Revised; NP, neutral position; E, extension; F, flexion; AR, axial rotation; LF, lateral flexion; IMUs, Inertial magneto units; ω_m, mean angular velocity; RMC, ratio of movement coupling; ICC, interclass correlation coefficient; P, participants

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small coupled lateral flexion to the same side occurs. Similarly, when lateral flexion is the primary movement performed, a small coupled ipsilateral rotation is observed (Panjabi et al., 1992). However, this mechanism is physiological and does not impede the intended movement. On the contrary, in the specific case of a weak neck in ALS, pure lateral-flexion is associated with a significant unwanted coupled out of plane axial rotation, which makes the intended movement more difficult (Pancani et al., 2017).

Patients living with Amyotrophic Lateral Sclerosis (plwALS) that experience neck muscle weakness are advised to wear a cervical orthosis to improve their posture, their ability to perform daily activities and ease discomfort. However, most commonly adopted cervical orthoses often provide inadequate support or are uncomfortable and are frequently rejected by the patients (Reed et al., 2015). The main limitation of those orthoses is that they have not been designed for the specific needs of plwALS (Reed et al., 2015). The Head Up collar (previously known as the Sheffield Support Snood) is a cervical orthosis which was designed in collaboration with plwALS to meet the needs of those with ALS who are affected by neck muscle weakness. The orthosis consists of a snood-like base, made of stretchable fabric. Onto the snood base a range of polypropylene supports can be attached through a Velcro hook and loop mechanism. The supports can be placed in any position to adapt the support offered, according to the task performed and the plwALS's level of functional limitation (Fig. 1) (Baxter et al., 2016). Furthermore, by varying the number and type of supports applied, the level of support offered during the progression of the disease can be changed.

Previous assessments on healthy individuals quantified objectively the restriction of movement and support the Head Up collar can provide. Of particular note was the ability to enable selected targeted head movements without limiting others (Pancani et al., 2016). Additionally, the mechanical support offered by the Head Up collar was comparable to that of more rigid and/or bulkier orthoses traditionally used by plwALS (Pancani et al., 2016). The acceptability of the Head Up has been evaluated, with plwALS reporting a number of benefits including the ability to provide support while allowing a satisfactory range of motion, flexibility of use, the appearance and the comfort offered (Baxter et al., 2016). However, the effectiveness of the Head Up collar in improving the amount and quality of the head movements in ALS patients has not yet been investigated quantitatively.

The aim of this study was to observe head movements in plwALS and to perform a quantitative evaluation of the effects of the Head Up. In particular, this study aimed to establish if the Head Up collar facilitated more controlled and less coupled movements of the head, without limiting the natural velocity at which movements are performed. As a direct investigation of the motion of the cervical spine could not be performed, movements of the neck were investigated through the assessment of the movements of the head with respect to the trunk.

2. Methods

2.1. Participants

Participants gave informed written consent prior to the participation in the study, which was approved by the local ethics committee (REC number STH18733). Recruitment was carried out for 18 months among plwALS attending the Sheffield ALS clinic Inclusion criteria were: definite diagnosis of ALS (modified El Escorial criteria (Brooks, 1994)) and neck muscle weakness as assessed by a physician (any neck muscle scoring less than MRC score 5, as assessed at the time of their first referral). Additional inclusion criteria were: ability to understand instructions and to perform the testing procedures. Individuals that were not able to raise their head from their chest in a sitting position were excluded from the study, as well as individuals for which a suitable size of the Head Up collar was not available (very small or large neck sizes). Thirteen plwALS were recruited and baseline characteristics are provided in Table 1. The same specialist nurse rated the

Fig. 1. The Head Up collar. a) From left to right: stretchable fabric snood, frontal Z-shape supports to be placed under the jaw, frontal A-shape support to be placed under the chin, straight support to be placed on the back of the neck, lateral support to be placed over the shoulder; b) Head Up collar with an A-shape support, frontal view.

Table 1
Participants' characteristics: age, gender, ALSFRS-R score (0–48) at the time of recording, months from the diagnosis, orthosis currently used, number and type of Head Up collar supports used, and head movements performed. All = participant performed: extension, flexion, axial rotation and lateral flexion. y = yes, n = no.

Participant (P)	Age (years)	Gender	ALSFRS-R score	Time from diagnosis (months)	Cervical orthosis currently used	Head Up collar: number and type of supports used	Head movements
1	69	F	30	11.5	Soft Orthosis	2 frontal Z-shape	All
2	74	M	13	49	NA	2 frontal Z-shape, 2 lateral	All
3	69	M	44	1.5	Headmaster	2 frontal Z-shape	All
4	63	F	18	36.5	NA	2 posterior	All
5	58	F	43	34.5	NA	2 frontal Z-shape	All
6	53	F	22	2.5	Headmaster	2 frontal Z-shape, 2 lateral	All
7	69	M	23	18	Soft Orthosis	2 frontal Z-shape	All
8	53	F	34	10	Soft Orthosis	no supports, only snood	All
9	65	M	19	36	Soft Orthosis	2 frontal Z-shape	All
10	74	M	17	59	Soft Orthosis	2 posterior	All
11	50	F	23	57	Soft Orthosis	2 frontal Z-shape	No flexion without orthosis
12	45	M	18	36	NA	2 frontal Z-shape	All
13	63	M	36	45	NA	No supports, only snood	All

severity of the disease using the ALS Functional Rating Scale - Revised (ALSFRS-R), a validated clinical scale ranging from 48 (best) to 0 (worst) (Cedarbaum et al., 1999).

2.2. Experimental protocol

Data were collected within a hospital setting (Royal Hallamshire Hospital, Sheffield, UK). Participants were asked to perform a series of active head movements while sitting on a chair (or in their own wheelchair). They were instructed to start from a neutral position (NP, maintaining an upright head position and looking forward), then perform an extension (E), a flexion (F), an axial rotation (AR, both on the left and right side) and a lateral flexion (LF, both on the left and right side) of the head, moving it as far away as possible from NP. Each of these movements can be considered as the combination of two phases: phase 1, from NP until the neck had reached the end of the possible range of movement and phase 2, from the end position back to NP. Those two phases were identified and analysed separately on the assumption that they involve different group of muscles.

Before the actual experiments, participants performed the whole range of active head movements at least once to familiarize themselves with the test procedure and to stretch the neck muscles. In the same session, they were fitted with a Head Up collar by a trained operator (over two years of experience), as per their individual need. The patients were first asked to indicate which area of the neck (frontal, posterior, lateral) they felt required support and to describe the movements they were limited in performing. The supports needed to sustain those areas and movements were applied to the snood base and patients were then asked to repeat whole range of active head movements wearing the Head Up collar. If needed, different configurations of the supports were tried, until the plwALS considered the Head Up collar was offering the support they preferred. The number and type of supports used by each participant are summarized in Table 1.

Participants were asked to perform three repetitions of each movement, both with and without the Head Up collar, if able to, otherwise to stop once they felt too tired to complete the task. The order in which the two batches of the three trials were performed by the patients was randomized. Trials with and without the Head Up collar were not mixed to reduce the burden to the patients associated with repeatedly taking-off of the Head Up and to avoid changes in its fitting.

Two Inertial magneto units (IMUs, OPAL, APDM Inc., USA, sampling frequency 128 samples/s) were firmly attached to the forehead and sternum of each participant, using double-sided tape. Each IMU uses a tri-axial accelerometer to measure the linear acceleration, a tri-axial gyroscope to measure the angular velocity and a tri-axial magnetometer to measure the sensor orientation. These have been demonstrated to be suitable for the assessment of neck kinematics and quantitatively assess neck functional limitation in patients with ALS (Pancani et al., 2017). The alignment of the two sensors reference frame was performed through a functional calibration procedure, extensively described in previous studies (Duc et al., 2013), during which the participants were required, while sitting, to perform a series of flexions of their trunk.

2.3. Data processing

All data were processed in MATLAB (R2015a, Mathworks Inc., Natick, MA, USA). Data were filtered using a 4th order zero-lag Butterworth filter with a cut-off frequency of 10 Hz (Pancani et al., 2017). The sternum accelerations and angular velocities were subtracted from those at the head, to identify and exclude from the analysis those movements of the head that were only a consequence of movements of the trunk (details available in Pancani et al. (2016, 2017)).

For each movement, the two phases 1 and 2 previously described were differentiated by detecting the instant when the angular velocity signal crossed the zero value, which coincides with the moment when the direction of a movement is reversed (see Fig. 2).

The mean angular velocity (ω_m) was calculated by averaging the signal recorded by the tri-axial gyroscope over the duration of the movement. The Ratio of Movement Coupling (RMC) was then calculated using the following equation (Pancani et al., 2017):

$$RMC = \frac{A_j + A_k}{A_i}, \quad (1)$$

where i is the axis perpendicular to the plane where the primary movement is performed, j and k are the other two main anatomical axes and A_i, A_j, and A_k are the areas under the angular velocity time-curves measured along those axes.

The RMC has been proven to be a viable parameter to quantify the presence of coupled movements (Pancani et al., 2017) in patients with ALS.

2.4. Statistical analysis

The repeatability of the ω_m and RMC values over the three trials was verified, for both conditions, by using a two-way random interclass correlation coefficient (ICC (2, 1)) for a single measurement (Shrout and Fleiss, 1979). According to the literature, ICC values were interpreted as: good > 0.75, moderate 0.4–0.75 and poor < 0.4 (Fleiss et al., 2003).

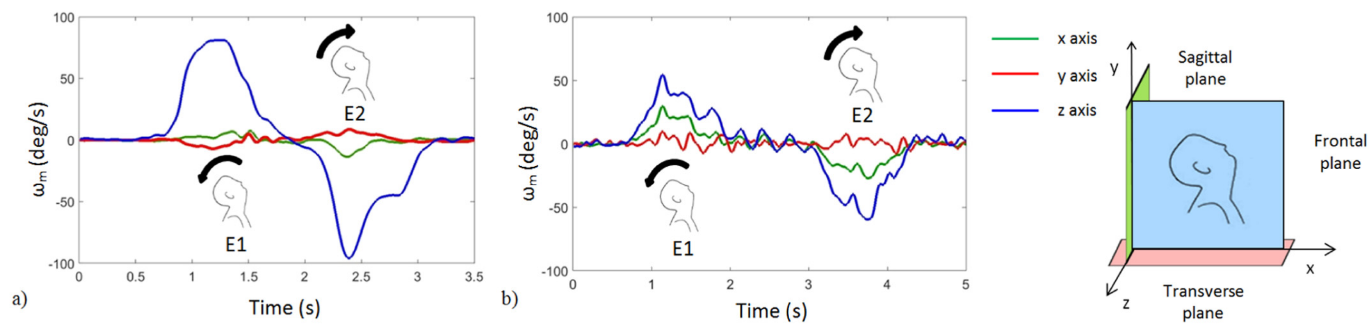

Fig. 2. Full extension movement (from and back to the Neutral Position) performed by a young healthy individual (a) and a patient with ALS (b). E1: movement from neutral position, E2: movement back to neutral position. 2a shows a predominance of the angular velocity along the z axis, consistent with the fact that the movement occurs entirely in the sagittal plane. In 2b, this predominance is less definite and components of the angular velocity along x and y axes are observed as a reflection of coupled movements occurring in the other anatomical planes.

To assess the effect of the orthosis, a first level of analysis was performed by averaging, for each movement, the values obtained in its three repetitions. Differences between the parameter measured with and without the Head Up collar were assessed by using a paired *t*-test or a Mann-Whitney *U* test, according to the normality or non-normality of data, as verified using a Shapiro-Wilk test. In both cases, statistical significance was set at an alpha level of 0.05. Cohen's d was calculated as an indicator of the effect size. The effect size was considered negligible when $d \le 0.2$, small when $0.2 < d \le 0.5$, medium when $0.5 < d \le 0.8$ and large when $d > 0.8$ (Cohen, 1977). A second level analysis was carried out by looking in detail at the RMC values measured during the three repetitions for each subject and for each movement.

3. Results

The group was characterized by a high variability in terms of age, time from diagnosis and progression of the disease (Table 1). The adopted configuration of the Head Up collar also differed among participants, ranging from a minimally restrictive (snood only) to a highly supportive setting (2 frontal and 2 lateral supports). The most frequently used supports were the two frontal Z-supports, chosen by 9 participants. ICC for the RMC was good in all movements except in LF1 when performed without orthosis and in AR2 and LF1 when performed with the Head Up collar (Table 2). For those movements, a moderate ICC was observed. Similarly, for ω_m, an ICC moderate to good was observed (Table 2).

Fig. 3 shows the angular velocity (ω_m) results. Since the data were normally distributed, the differences between movements performed with and without the Head Up collar were assessed using a *t*-test, using the average value among the three repetitions of each movement. As shown in the graph, no evident trend could be observed, and no differences were found between the two groups.

Fig. 4 shows the average value of RMC among three repetitions, for movements performed with and without the Head Up collar. Mann-Whitney *U* test was used for comparison since data were not normally distributed. Outliers, displayed as circles in the graph, are those values between 1.5 and 3 times the interquartile range, represented by the high of the box. Extreme outliers, displayed as stars, represent cases that have values more than three times the interquartile range. In Fig. 4, labels associated with outliers and extreme outliers, hence indicate those participants that showed RMC values distant from the median of the observed sample. A reduction in the number of outliers, and a consequent reduction in RMC values for those that showed the higher impairment in performing the movement, was observed in all movements executed with the Head Up, except in AR. A significant reduction of coupled movements was observed in LF back to the neutral position (LF2, $P = 0.013$, $d = 0.72$). Comparison between other movements showed a small (F1: $d = 0.20$, F2: $d = 0.26$, AR1: $d = 0.22$, LF1: $d = 0.30$) or negligible (E1: $d = 0.18$, E2: $d = 0.16$, AR2: $d = 0.1$) effect size.

Fig. 5 details the effects of the use of specific supports, showing the RMC values measured with and without the Head Up collar only for those participants (P) who had their orthosis fitted with two frontal Z-shape supports (i.e. the most common configuration). These two supports are expected to affect only the frontal and lateral flexions: the Z-shape supports are expected to sustain and slow down the head when moving forward by pulling it from the jaw, and to help it to rise from a frontal flexion by offering a push. Similarly, when performing LF1 and LF2 the Z-shape supports are expected to work as footholds, sustaining the weight of the head and reducing the load to be compensated by the neck muscles. Not all patients managed to perform three repetitions of each movement, mainly due to excessive fatigue. In particular, patients 1, 9 and 11 were able to execute only two repetitions of the F1 movement while wearing the Head Up collar. Patients 7 and 9 experienced excessive fatigue and were only able to complete 2 repetitions of all F2 tasks.

The use of the Head Up collar was beneficial for P5 and P12, for whom out of plane movements were reduced when performing F1. Similarly, P12 showed improved control of the head movement also in performing F2. The positive support to flexion offered by the Head Up collar was evident for P11, who was able to perform both F1 and F2 only when wearing it. Only P1, on the contrary, had higher RMC, and hence worse head control, in both F1 and F2 when wearing the Head Up.

Concerning LF, a lower RMC was found in P9 and P12 when performing LF1 with the Head Up collar, while a higher value was observed in P7 in the same condition. Finally, P9, P11 and P12 showed an improvement toward the reduction of out of plane movements, when performing LF2 with the Head Up.

Table 2
ICC values for the ratio of movement coupling (RMC) and mean angular velocity (ω_m) measured without and with the Head Up collar in the extension (E) flexion (F), axial rotation (AR) and lateral flexion (LF) movements. 1: movement from neutral position; 2: movement back to neutral position.

ICC	RMC		ω_m	
	Without Head Up collar	With Head Up collar	Without Head Up collar	With Head Up collar
E1	0.85	0.78	0.57	0.72
E2	0.88	0.88	0.66	0.52
F1	0.95	0.92	0.65	0.87
F2	0.97	0.94	0.59	0.55
AR1	0.92	0.92	0.61	0.76
AR2	0.83	0.69	0.69	0.71
LF1	0.68	0.57	0.69	0.80
LF2	0.83	0.78	0.87	0.80

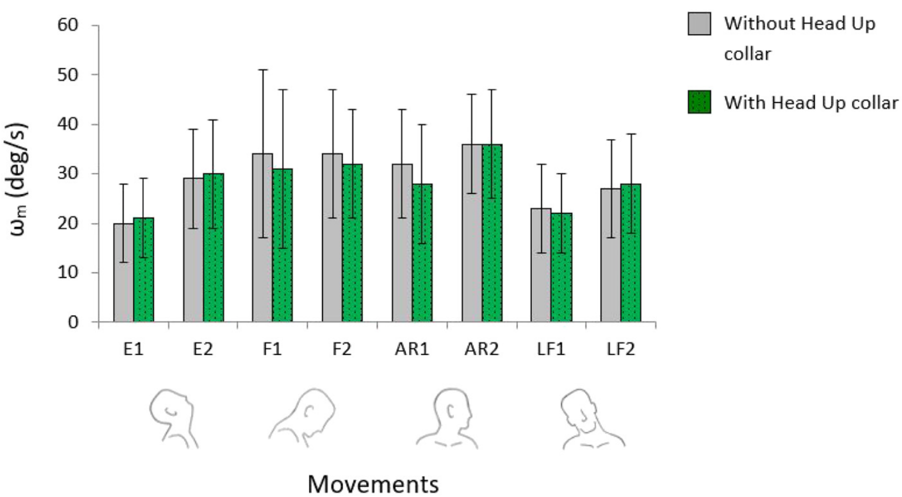

Fig. 3. Mean angular velocity (ω_m) values measured in the extension (E) flexion (F), axial rotation (AR) and lateral flexion (LF) movements (1: movement from neutral position; 2: movement back to neutral position) when performed without and with the Head Up collar. Values are presented through their mean and standard deviation. Statistical comparison is not reported since the differences were never significant.

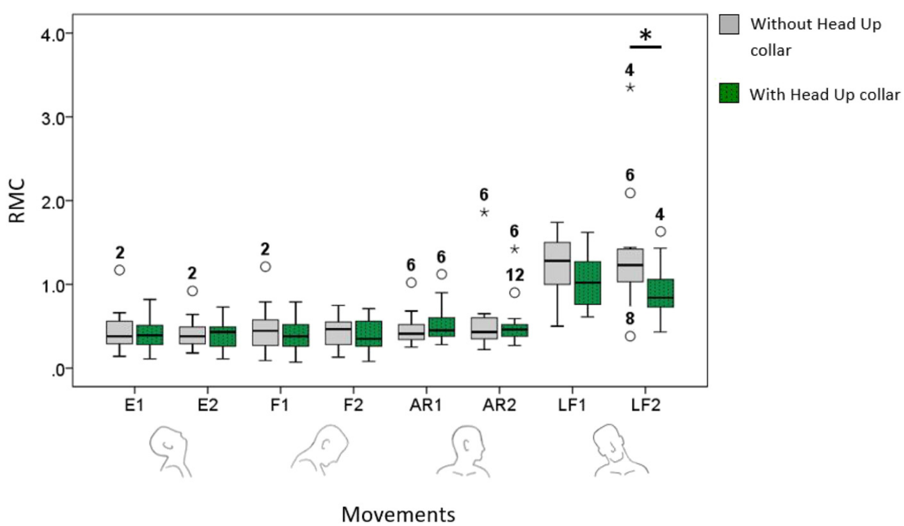

Fig. 4. Ratio of movement coupling (RMC) values measured in the extension (E) flexion (F), axial rotation (AR) and lateral flexion (LF) movements (1: movement from neutral position; 2: movement back to neutral position) when performed without and with the Head Up collar. Values are presented through the median, upper and lower quartiles and upper and lower extremes. Outliers and extreme outliers are represented by circle and stars, respectively. Number above the outlier indicates the patient associated to that value. (*) Level of significance for the difference between trials performed without and with the Head Up collar < 0.05.

4. Discussion

Poor control of active head movements with the presence of coupled movements has recently been described in plwALS (Pancani et al., 2017). The aim of this study was to verify whether the use of the Head Up collar, specifically designed for plwALS, could compensate for this poor control without limiting the natural movement velocity. This was achieved through a quantitative assessment.

Even if preliminary, the results achieved through the quantitative functional evaluation approach proposed in this study are certainly encouraging. Participants' movements exhibited a better repeatability in terms of RMC compared to movement velocity. Experimental measurements obtained highlight and quantify the heterogeneity that characterized the participants who exhibited different RMC values when performing the head movements, and diverse levels of impairment were observed for the same subject in performing different movements. An example is represented by P11: although having a good control of the movement in LF (Fig. 5c and d), P11 was not able to perform F without being supported by the Head Up collar (Fig. 5a and b). This heterogeneity in muscle weakness and functional compromise is consistent with that described more generally within the wider ALS population (McDermott and Shaw, 2008). It also reinforces the need for personalized interventions, aimed at offering support, according to the specific needs of the individual. The assessment approach adopted in this study provided repeatable and reliable information about the execution of head movements. This quantitative information may be used to unveil important characteristics of the patients' movement strategy, which might not be caught by a traditional qualitative assessment. This paves the way for the development of clinical evaluation methods aimed at monitoring the disease progression and/or assessing the effectiveness of an intervention. Understanding which movements are impaired (for example frontal flexion rather than extension) and to what extent, may guide the choice of a cervical orthosis or, in the case of the Head Up collar, of the supports needed for a more effective and patient specific fit.

The natural velocity of the movements was not affected by the Head Up collar; with the angular velocity remaining as high as when the movements were performed without it. This can be certainly regarded as a positive result as in general the velocity has a significant functional relevance and its decline is normally undesirable.

The quantification of the movement coupling (RMC) demonstrated that the major improvement associated with wearing the Head Up was in the control of LF movement when returning to NP. The positive impact of the Head Up collar on this movement was likely generated by the frontal supports used, characterized by a “Z” shape and attached below the jaw (Fig. 1a). These supports were designed to sustain and guide the head in performing a frontal flexion while offering a lateral support base, below the jaw, that facilitates the lateral flexion. Furthermore, a reduction in the number of outliers was observed in movements executed with the Head Up collar, as can be observed for patient 2 in performing E1, E2 and F1 and for patient 6 in performing LF2, indicating that those patients who started from a higher level of

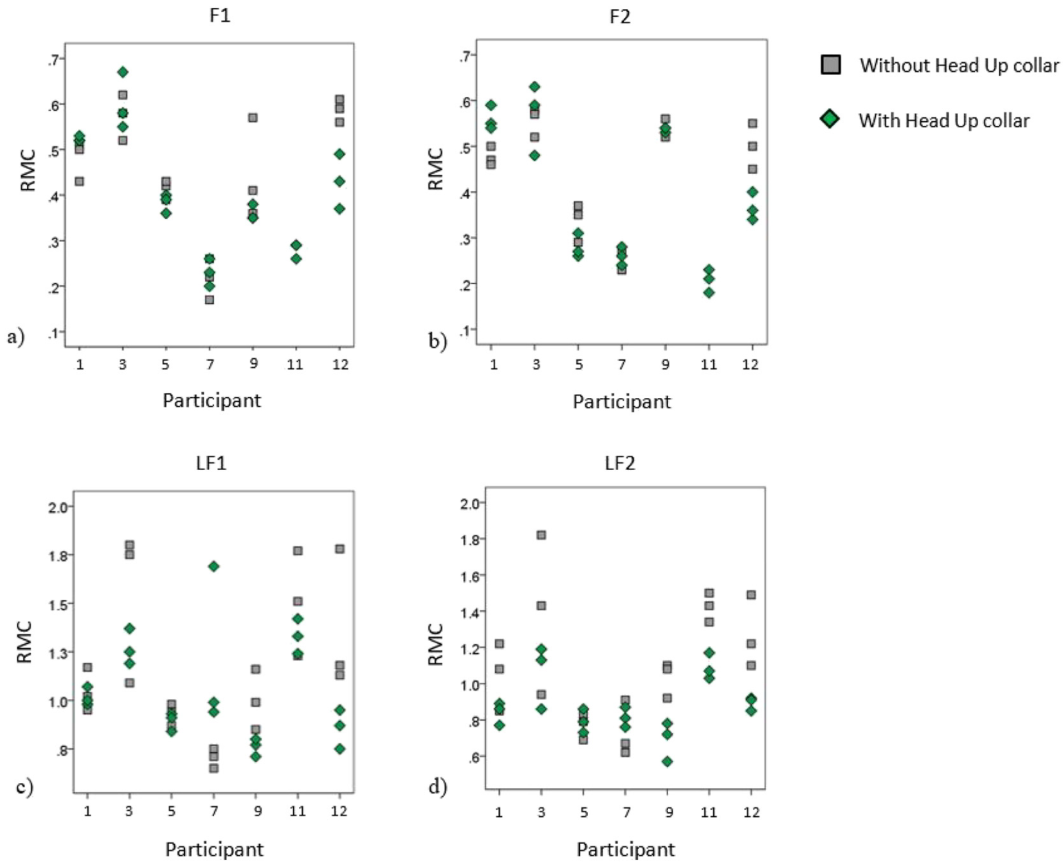

Fig. 5. RMC values measured in the three trials performed without orthosis (grey squares) and the three trials performed with the Head Up collar with two frontal Z-shape supports (green diamonds). Movements reported are Flexion (F): from neutral position (a) and back to neutral position (d) and Lateral Flexion (LF): from neutral position (c) and back to neutral position (d). (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

impairment were also those who benefitted the most from wearing the collar. As the fitting of the Head Up was based only on the patients' preference and feedback some participants were given supports that they might not have really needed. P7, for example, requested frontal supports although his ability to perform F and LF movements was not compromised (Fig. 5). This observation was confirmed also for other configurations and movements: P4 requested posterior supports, although her results indicated that she was significantly impaired in performing LF (Fig. 4) and might have benefited more from the use of supports placed under the jaw. Finally, P2 did not ask for posterior supports, although he exhibited poor control when performing E (Fig. 4). These results and considerations clearly indicate that a fitting of the Head Up collar based on a combination of a quantitative functional assessment of the patients and their preferences would likely further improve the efficacy of the intervention. Additional studies are of course needed to verify this hypothesis.

The reported results, despite having been obtained from a relatively small number of participants, which represents a limitation of this work, are encouraging in relation to the use of the Head Up collar in plwALS affected by neck muscle weakness. The absence of a randomization in the order of the trials might represent a limitation of this study, since plwALS easily experience muscular fatigue. This may have affected the trials with the collars. Further work is needed to verify this hypothesis. A further limitation of this study is the lack of a detailed evaluation of neck muscle strength. At screening, a brief assessment was performed to identify if at least one muscle exhibited weakness (MRC score < 5). A more comprehensive muscle testing would enable an informative comparison with data obtained from inertial sensors and is an aim for future work.

In addition, we asked the participants to wear the Head Up collar for about 30 min, which corresponds to the time required to complete the protocol, since we were interested in assessing the immediate response to the intervention. Reported results therefore do not account for possible discomfort or changes in the response coming from wearing the orthosis for an extended amount of time, which was evaluated in a previous study (Baxter et al., 2016).

5. Conclusions

In this preliminary evaluation, the Head Up collar was effective in enabling more controlled head movements for plwALS. As per any orthosis, a key factor for the effectiveness of the Head Up collar appears to be the need for a fitting based on the functional assessment of the patients in addition to their preference. After evaluating which movements are impaired and to what extent, an informed and objective decision can be made about the choice of the orthosis and its configuration. The chosen functional assessment parameters (RMC and angular velocity) have been shown to be valuable in assessing the functional limitations of neck movement and in evaluating the benefit of an orthoses. This approach may have value when applied to other areas of the body to evaluate impairment and subsequently effectiveness of any intervention which aims to improve the efficiency of a movement.

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