The effects of facioscapulohumeral dystrophy and dynamic arm support on upper extremity muscle coordination in functional tasks

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ABSTRACT

This study’s objective is to understand the effect of muscular weakness in persons with facioscapulohumeral dystrophy as well as the effect of a dynamic arm support on muscle coordination and activity performance, during activities of daily living. People with facioscapulohumeral dystrophy (n=12, 56.0±14.5 years) and healthy controls (n=12, 55.5±13.4 years) performed five simulated daily activity tasks, while unsupported and supported by the Gowing dynamic arm support. Surface electromyography, kinematics, and maximum force output were recorded. Outcomes were calculated for muscle coordination (muscle synergies), maximum muscle activity, movement performance indicators, and upper limb muscular weakness (maximum force output). Muscle coordination was altered and less consistent in persons with facioscapulohumeral dystrophy compared with healthy controls. The dynamic arm support alleviated muscle efforts and affected muscle coordination in both populations. While populations became more similar, the internal consistency of persons with facioscapulohumeral dystrophy remained unaffected and lower than that of healthy controls. Furthermore, the support affected movements’ performance in both groups. The maximum force outputs were lower in persons with facioscapulohumeral dystrophy than controls. Muscle coordination differences were presumably the result of individual-specific in muscle weakness and compensatory strategies for dealing with gravity compensation and movement constraints.

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1. Introduction

Facioscapulohumeral dystrophy (FSHD) is considered one of the most prevalent neuromuscular disorders with an estimated two thousand affected individuals in The Netherlands in 2010 [1]. FSHD is characterized by progressive loss of muscle strength, mostly in the shoulder area, increased fatigue, pain, and joint stiffness [2–6]. People with FSHD have difficulties performing activities of daily life (ADL) and often show compensatory strategies requiring increased effort and energy [7]. Dynamic arm support devices compensate for gravity and consequently improve ADL performance for people with muscular weakness [8,9]. However, Heide et al. showed that a discontinuous use of dynamic arm support devices is reported in the majority of user studies [8], which implies a suboptimal use of these devices. A better understanding of how dynamic arm support devices influence body functions and ADL may contribute to their further development and increase usage rate [10].

Muscle weakness of the shoulder girdle significantly limits the ability of people with FSHD to perform independent ADL [11,12]. Bergsma et al. showed that eating, drinking, and reaching are severely limited in these persons, with ~42% experiencing extreme difficulties to reach forward at shoulder level and ~80% to reach over their head [3]. These limited activities are generally accompanied by increased muscle activities of biceps, deltoid, trapezius, and pectoralis muscles, which are ~3–5 times higher than in healthy individuals [7]. In addition, FSHD affects muscle coordination of the shoulder girdle during arm lifting, resulting in a reduced contribution to scapular upward rotation by the trapezius ascendens and serratus anterior up to 41% [13,14]. Typically, a dynamic arm support provides an enhanced ability to reach and repeatedly lift the arm during ADL such as personal care, eating, and drinking [9,15–17]. However, a dynamic arm support can also induce mechanical constraints resulting in longer movement time and altered smoothness [16,18–20]. The support device may thus influence shoulder muscle coordination,
potentially leading to a destabilizing effect on the glenohumeral joint. This can occur as the upward force imposed by the device may demand a greater effort by the glenohumeral joint muscles, thus also leading to long-term fatigue. Therefore, examining the effects of muscular weakness and adaptations that may occur from the use of an arm support device during ADL is important to understand the long-term implications of such devices.

In the current study we use muscle synergy analysis to quantify the changes in muscle coordination while using a dynamic arm support. Research has shown that the central nervous system controls groups of synergistic muscles to solve the motor redundancy problem [21]. Muscle synergy analysis simplifies the representation of muscle coordination patterns to a lower dimensional spatiotemporal output of synergistic contributions (weights) and activation patterns (coefficients) [21]. Four parameters are commonly used to quantify the variability and alterations in muscle coordination: 1) the number of muscle synergies required, 2) the variances accounted for per synergy, 3) synergy similarities between groups or conditions, 4) and synergy consistency within the same group or condition [13,16,19,22-28].

It has been shown that dynamic arm support devices have little influence on the muscle coordination of healthy (older) participants, regardless of the level of support [16,19,25]. In people with FSHD, it can be speculated that an arm support would alter the selection of synergistic shoulder elevation muscles over time in a more pronounced way than in healthy persons. Moreover, overcoming the additional external force resulting from the gravity compensation device, could require altered synergies compared to healthy persons, due to the muscle weakness of the arm adductors in persons with FSHD. Movement performance, e.g. task duration and movement smoothness would consequently also be affected. However, muscle synergies in FSHD persons using a support remain unclear at present.

Altered muscle coordination patterns in persons with FSHD, following the use of an arm support device, may influence factors such as fatigue or susceptibility to injury, which are likely to influence usage rates. Knowledge of how muscular weakness and arm support devices influence muscle coordination and activation is needed for the continued development of such devices. Therefore, the aim of this study is to investigate the effect of muscular weakness in persons with FSHD, as well as the effect of a dynamic arm support on muscle coordination and activity during ADL tasks [10]. Furthermore, the effect of the dynamic arm support on the movement execution is quantified. Our primary hypothesis is that, muscle coordination when performing ADL without the arm support is less consistent within the FSHD group than within the healthy control group and is influenced by the type of ADL performed. Our secondary hypothesis is that using a dynamic arm support results in a more consistent muscle coordination, with a larger increase in consistency within the FSHD group compared to the healthy control group. Thirdly, we also hypothesize that using the arm support would lead to more similar synergies, i.e. muscle coordination would become more similar between the two groups.

2. Material and methods

2.1. Participant characteristics and inclusion criteria

Data were collected from participants with FSHD and healthy controls in a larger study approved by the central Medical Ethical Committee of the University Medical Center Groningen (NL5711.042.15). The study was conducted in accordance with the guidelines of the Helsinki protocol. Participants were aged between 18-75 years, able to read and understand Dutch, and able to give written informed consent. Additionally, people with FSHD were included if they were able to transfer from a wheelchair to a chair (including with manual assistance), and had a Brooke scale score of 3 or 4. Healthy participants were excluded if they had any pathologies, shoulder pain, or a history of severe trauma of the shoulder <2yrs (e.g. fracture, luxation) that could interfere with the measurement results. Exclusions criteria for participants with FSHD were as follows: comorbidities that could interfere with the measurement results, previous surgery on the right shoulder, extrinsic causes of shoulder pain, a history of severe shoulder trauma, or an inability to elevate the right arm above 30°.

2.2. Tasks

The participants were seated in a chair with a left side-rest and lower back-rest and with the seat height set to achieve a knee flexion angle of 90°. Participants received detailed instructions regarding the movement before the execution of each task. Five tasks were chosen, according to the categories provided by Bergsma et al. [3], to reflect a selection of important ADL. Tasks were repeated three times in a randomized order. The tasks included 1) pushing and pulling (PP) an object, 2) simulated drinking with a cup of 200 grams (C2M), 3) simulated eating with a spoon (S2M), and 4) reaching towards a target at shoulder height on the ipsilateral side (ILR) and 5) on the contralateral side (CLR). Tasks PP, ILR, and CLR were performed at one shoulder width from the participant’s midline to the respective side. Participants were allowed to rest for a few minutes between tasks and repetitions. All tasks and repetitions were first completed without the dynamic arm support and followed by a rest period of fifteen minutes. Successively, tasks were once more randomized and performed with the dynamic arm support. This sequence of task execution wo/w the device and incorporation of resting periods were to minimize fatigue and ensure protocol completion.

2.3. Dynamic arm support

The Gowing dynamic arm support (Focal Meditech BV, Tilburg, Netherlands) (Fig. 1) provides spring-actuated passive support at the lower arm, where the spring tension is adjustable by motorized actuators [29]. The amount of support was personalized to simulate a gravity-free sensation and was constant within the reachable task workspace. Participants had no previous experience.

![Fig. 1. A participant with FSHD performing the push and pull task with the Gowing viewed from posterior (left) and lateral (right) perspective.](image)
with a dynamic arm support device and were given up to 10 minutes of familiarization time prior to performing the tasks.

2.4. Measurement and processing

Kinematics of the right hand, using an active marker placed on head of the 3rd metacarpal, were recorded at 100Hz using the Optotrack 3020 system and NDI First Principles application (Northern Digital Inc., Canada) [30] and used to calculate movement performance as in task duration, smoothness, and efficiency (see Section 2.6 Kinematics). Surface electromyograms (EMG) were recorded for muscles on the right side, which included the prime humeral elevator/depressors and scapular rotator muscles, i.e. medial deltoid, pectoralis major clavicular head, latissimus dorsi, trapezius descendens, trapezius ascendens, and serratus anterior 5-6th rib, and the synergist muscles biceps brachii short head and triceps brachii long head. Data were captured at 2000Hz using the Delsys Trigno Wireless EMG system and EMGworks Acquisition application [31]. Skin was prepared and sensors were placed according to the Surface Electromyography for the Non-Invasive Assessment of Muscles (SENIAM) guidelines [32].

Maximum voluntary contractions (MVCs) during isometric conditions were recorded beforehand (Appendix Table 1). The recorded EMG data were filtered with a 4th order Butterworth 20-450Hz bandpass and a 49-51Hz bandstop filter, rectified, smoothed with a 100ms moving window, normalized to the maximum amplitudes derived from all MVC and task recordings, and filtered with a 4th order Butterworth 5Hz low pass filter. The maximum task-specific muscle activity was extracted as highest normalized amplitude over all task repetitions. Time was normalized to 1001 samples for each repetition ranging from 0 to 100%.

In addition, force output of the shoulder elevators, humeral elevators, and elbow flexors were measured with a load cell, AST KAP-S/KAP-E Force Transducer [33], at 100Hz during the MVC recordings to evaluate muscle strength in the two groups. The load cell was attached to the chair to minimize the burden on the participants in terms of transfers and time. One researcher provided instructions to ensure the correct position and execution (Appendix Table 1) of respective isometric contractions for shoulder elevation (during the trapezius descendens recording), humeral elevation (during the medial deltoid recording), and elbow flexion (during the biceps brachii recording). Participants were instructed and encouraged to contract maximally for five seconds, which was repeated after two minutes rest. The force output was visually checked and extracted as the maximum force during these five seconds of both repetitions.

2.5. Muscle synergy extraction

EMG data were pooled for task repetitions per individual in a single matrix to investigate the muscle synergies between the two groups and two support conditions within respective tasks. Muscle synergies were extracted using non-negative matrix factorization, which decomposed the matrix into 1 to 8 sets of components consisting of weights and coefficients [21]. The weights and coefficients were converted to a unit vector and represent normalized muscle activity (0-1). Furthermore, the non-negative matrix factorization provided the percentage of variance accounted for of all muscles and per individual muscle for each set of synergies. At least 90% of all muscles’ and >75% of individual muscles’ variance should be accounted for before a set of synergies was considered to represent muscle coordination. The synergies were then clustered based on the Pearson’s correlation coefficients (r) calculated between all possible combinations of individual participants’ synergy weights within respective group, support condition, and task [28,34]. Clustered synergies, which represented the muscle coordination of a group, for a support condition and a task, were then ranked in an ascending order (MS1-4) based on the number of participants in each cluster.

Subsequently, in a leave-one-out process [26,27] synergy consistency, which refers to correlations within clustered synergies, was calculated as the Pearson’s correlation coefficients between the synergy weights of individual participants included in the cluster and the mean synergy weights of the cluster without that given participant. Furthermore, synergy similarity, which refers to correlations between clustered synergies, was calculated as the Pearson’s correlation coefficients between the synergy weights of individual participants of one cluster and the mean synergy weights of another cluster within respective tasks [34]. For similarity calculations between groups, FSHD individuals were compared with the mean of controls, and between support conditions, individual synergy weights while supported were compared with the mean while unsupported. Furthermore, the synergy consistency and similarity calculations were restricted to 1) equally-ranked clustered synergies and 2) the first (MS1) and second (MS2) ranked synergies, since these account for the majority of the EMG variance, which were on average >50% and >33%, respectively.

2.6. Kinematics

Indicators of movement performance, as in task duration, smoothness, and efficiency, were calculated similarly as in Pirodini et al. [19]. Task duration was calculated as time in seconds between start and end of movement. Smoothness was calculated as the median jerk of the finger marker. Efficiency was calculated as the root mean square error between the trajectory of the finger marker and straight lines between start, target, and end, and normalized for length where i represents one sample and n represents the entire data set:

\[
\text{efficiency} = \frac{\left( \sum_{i=1}^{n} \text{(finger trajectory - straight line)}^2 \right)}{\sqrt{\sum_{i=1}^{n} \text{(straight line)}^2}}
\]

The start, target, and end positions were determined as the respective positions where velocity was closest to zero. Task duration, smoothness, and efficiency values towards zero represent a fast, smooth, and efficient movement, respectively.

2.7. Statistical analysis

Ten parameters were extracted to investigate the effect of FSHD and a dynamic arm support on muscle coordination with respect to movement performance (Table 1).

Our first hypothesis is that synergies in the FSHD group are less consistent than in the control group. Our second hypothesis is that a dynamic arm support results in more consistency, thus the synergy consistency of supported tasks should be higher than unsupported tasks. To test these two hypotheses, a non-parametric analysis of variance [35] was performed on the synergy consistency of the first and second ranked synergies, with population as between group factor and support conditions and tasks as within group factor (α = 0.05). The Pearson’s correlation coefficients were first transformed with the Fisher’s z-transformation formula [36] to normalize the sampling distribution:

\[
\text{Fisher’s z-transformation} = 0.5 \times \ln \left( \frac{1 + r}{1 - r} \right)
\]

In addition, Cohen’s d [37] was calculated from the Pearson’s correlation coefficients of the groups’ mean (M) and standard
deviations for both groups (SD):

\[
Cohen's \ d = \left( \frac{(M2 - M1)}{SDT} \right) \quad \text{(3)}
\]

The Cohen's d was then corrected with the unbiased d formula as Hedges' g [38], which includes sample sizes for the correction (df):

\[
Hedges' g = Cohen's \ d \times \left( 1 - \left( \frac{3}{4 + \frac{df}{2}} \right) \right) \quad \text{(4)}
\]

The Hedges' g (Hg) was interpreted as very small (0.00- 0.01), small (0.01 - 0.20), medium (0.20 - 0.50), large (0.50 - 0.80), very large (0.80 - 1.20), and huge (>1.20) [13,37].

Post-hoc analyses were performed for significant effects in the analyses of variance, with the exception of task effects, as a Wilcoxon signed rank test for related samples and Wilcoxon rank sum test for unrelated samples. Tasks were pooled and alpha levels were corrected accordingly using the Bonferroni method and set to 0.01.

To test our third hypothesis, that muscle coordination would become more similar between the two groups under the influence of an arm support, differences in muscle synergy similarity were investigated with a Wilcoxon rank sum test after the Fisher's z-transformation (formula 2). First, we compared the similarities calculated between the unsupported FSHDs and supported controls with the similarities calculated between the supported FSHDs and supported controls to test the effect of support. Second, we compared the similarities calculated between the unsupported FSHDs and unsupported controls with the similarities calculated between the supported FSHDs and unsupported controls to test the interaction effect of support and FSHD. Comparisons were performed respectively on the first and second ranked synergies. The Hedges' g was calculated as a range of effect size. Tasks were pooled and alpha levels were corrected accordingly using the Bonferroni method and set to 0.01.

Additional analysis was performed on the secondary outcome measures (Table 1). The Hedges' g was calculated as a range of effect size for all supplementary analyses. In all analyses, tasks were pooled and alpha levels were corrected accordingly using the Bonferroni method and set to 0.01.

First, the number of extracted synergies were compared between FSHD and control group for support conditions with a Wilcoxon rank sum test and subsequently between support conditions respectively for the FSHD and control group with a Wilcoxon signed rank test.

Second, to evaluate the limitations of muscular weakness, the maximum muscle activity during tasks was tested with a non-parametric analysis of variance with population as between group factor and support conditions and tasks as within group factors (\(\alpha = 0.05\)). Post-hoc analyses were performed for significant effects in the analyses of variance, except for task effects, as a Wilcoxon signed rank test for related samples and Wilcoxon rank sum test for unrelated samples.

Third, to investigate the effect on task performance, a non-parametric analysis of variance was performed on the task duration, jerk, and efficiency (\(\alpha = 0.05\)). Post-hoc analyses were performed for significant effects in the analyses of variance, except for task effects, as a Wilcoxon signed rank test for related samples and Wilcoxon rank sum test for unrelated samples.

Fourth, to quantify muscular weakness the maximum voluntary force output during shoulder elevation, humeral elevation, and elbow flexion were compared between populations with a Wilcoxon rank sum test (\(\alpha = 0.05\)).

3. Results

3.1. Participant characteristics

Twelve healthy control participants (6M/6F, 55.5 ± 13.4yrs, 1.76 ± 0.08m, 72 ± 14kg, 11 right- & 1 left-handed) and twelve participants with FSHD (6M/6F, 56.0 ± 14.5yrs, 1.76 ± 0.10m, 75 ± 20kg, 9 right- & 3 left-handed) were included in this study.

3.2. Muscle synergies

Up to four syneriges were extracted per task where more than 70% of the participants generally required two synergies to perform a task (Appendix Figure 1). Synergies were less on average for the FSHDs than for the controls (unsupported p: 0.002, Hg: -0.59 to -0.49, and supported p: 0.003, Hg: -0.57 to -0.50) (Appendix Figure 1). The number of extracted synergies could not be found to differ per participant between support conditions. The clustered synergy weights for the contralateral reaching task are shown in Fig. 2, while weights and coefficients for other tasks are shown in the appendix (Appendix Figures 2-10).

In the unsupported contralateral reaching task (Fig. 2 and Appendix Figure 10), the controls' first ranked synergy involves scapular mobility and stabilization, mostly by the trapezius. In the second ranked synergy, similar functions are present, in particular upward scapular rotation by the trapezius descendens and serratus anterior and were more actively accompanied by up- and inward rotation and stabilization of the humerus by the deltoid, pectoralis, and latissimus dorsi, respectively. In FSHDs, the first ranked synergy resembles a merge of the controls' first and second ranked synergies. Their second ranked synergy represents a co-contraction that involves elbow flexion, scapular downward rotation, and humerus depression, mostly by the biceps, trapezius ascendens, and latissimus, respectively. In the supported contralateral reaching task, minor differences can be noted in controls while FSHDs present a shift in deltoid and trapezius ascendens contributions between the first and second ranked syneriges.

Table 1

Overview of study outcome parameters with input data, conversion method, and unit.

| Biomechanical characteristic | Input data | Outcome parameter | Method | Unit |
|-----------------------------|------------|-------------------|--------|------|
| Muscle coordination         | EMG        | Number of syneriges | Non-negative matrix factorization | # |
|                             |            | Synergy weight consistency | Pearson correlation coefficients within clustered synergies | r |
| Muscle activity             | EMG        | Muscle activity | | |
| Movement                     | Kinematics | Task duration | Pearson correlation coefficients between clustered synergies | 0-1 MVC |
| performance                  |            | Smoothness | | seconds |
| Muscular weakness           | Kinetics   | Efficiency | Jerk | mm/s~1 |
|                             |            | Shoulder elevation strength | Root Mean Square Error | mm |
|                             |            | Humeral elevation strength | Maximum force output | N |
|                             |            | Elbow flexion strength | Maximum force output | N |

Numbers represent the primary (1) and secondary (2) outcome parameters. EMG: ElectroMyoGrams, MVC: Maximum Voluntary Contraction.
3.3. Synergy consistency

A significant group effect for the weight consistency was found for MS1 and MS2, with FSHDs less consistent than controls in MS1 (p<0.001, task averaged Pearson’s correlation coefficients r: -0.34, Hg: -1.48 to -0.98) and MS2 (p<0.001, r: -0.41, Hg: -1.12 to -1.08). In addition, a significant support effect was found for MS1, where consistency was generally higher in the supported tasks (p<0.001, r: +0.14, Hg: 0.41 to 0.47).

Furthermore, a significant group * support interaction effect was found for MS2, but not MS1 (p: 0.110), with FSHD less consistent than controls. For unsupported movements the difference in consistency between FSHDs and controls was r: -0.26 (p<0.001, Hg: -0.69 to -0.65) and for supported movements r: -0.54 (p<0.001, Hg: -1.61 to -1.60). Between supported FSHD and unsupported controls the difference was r: -0.39 (p<0.001, Hg: -1.15 to -0.96) and for unsupported FSHD and supported controls r: -0.42 (p<0.001, Hg: -1.24 to -1.08).

Moreover, a significant group * support * task interaction effect was found for MS1, while for MS2 the group * support interaction effect (p: 0.002), as explained above, and the support * task interaction effect (p: 0.009) were significant. Post-hoc analyses showed that MS1 of unsupported FSHDs was significantly less consistent than unsupported controls (Push and Pull, p<0.002, r: -0.40, Hg: -1.64 to -1.31, Spoon to Mouth, p: 0.006, r: -0.36, Hg: -1.89 to -1.04) (Fig. 3A), but there was no difference between supported FSHDs and unsupported controls. For MS2, there were no group differences while both were unsupported (Fig. 3B), but supported FSHDs were significantly less consistent than unsupported controls (Push and Pull, p<0.001, r: -0.74, Hg: -3.55 to -2.32, Contralateral Reaching, p: 0.004, r: -0.39, Hg: -1.06 to -0.97). Furthermore, controls showed a significant increase in consistency from without support to with support in MS1 (Ipsilateral Reaching, p<0.001, r: +0.28, Hg: 1.03 to 4.92, Contralateral Reaching, p: 0.002, r: +0.25, Hg: 0.91 to 4.23) (Fig. 3C) and in MS2 (Spoon to Mouth, p: 0.006, r: +0.40, Hg: 1.02 to 2.89, Ipsilateral Reaching, p: 0.002, r: +0.36, Hg: 0.83 to 1.21) (Fig. 3D). There were no significant effects of support in the FSHD group. Additionally, significant task effects (MS1 and MS2) were found.

3.4. Synergy similarity

In unsupported movements it was found that the task averaged similarity between individual FSHDs and the mean of controls was r: 0.19 for MS1 and r: 0.07 for MS2 (Fig. 4A-B). The similarity between the synergies without and with the support were r: 0.23 (MS1) and r: 0.00 (MS2) for FSHDs and r: 0.72 (MS1) and r: 0.52 (MS2) for controls (Appendix Figure 11). Furthermore, the similarity between FSHDs and controls significantly increased when FSHDs used a support while controls were unsupported for MS1 (p<0.001, r: +0.12, Hg: 0.59 to 0.65), but not for MS2 (p: 0.454, r: +0.03, Hg: 0.16 to 0.16) (Fig. 4C-D). Finally, the similarity between FSHDs and controls significantly increased when both groups used a support compared with when both groups did not use a support for MS1 (p: 0.008, r: +0.12, Hg: 0.48 to 0.64), but not for MS2 (p: 0.409, r: +0.04, Hg: 0.17 to 0.19).

**Fig. 2.** Clustered muscle synergy weights during contralateral reaching for control without support (white, WO_S), control with support (light gray, WI_S), FSHD without support (gray, WO_S), and FSHD with support (dark gray, WI_S) ranked horizontally in order of prevalence (N). Bars represent the mean amplitude and lines one standard deviation.
3.5. Maximum muscle activity

A significant group effect was found for the maximum muscle activities of the biceps, deltoid, pectoralis, and latissimus, while a significant support effect was found for biceps, triceps, serratus, and latissimus. Finally, a significant group * support * task interaction effect was found for trapezius descendens, serratus, and latissimus (all p<0.001) with amplitudes of serratus and latissimus lower in controls in selected tasks with the support. Details are reported in the Appendix (appendix figures 12-13). In addition, all muscles, except for pectoralis and serratus, presented a significant task effect and the majority of muscles a significant support * task interaction effect.

3.6. Force output

The maximum voluntary force output was significantly lower in FSHDs than controls for shoulder elevation (p: 0.002, -166N and Hg: -1.82 to -1.21), humeral elevation (p: 0.032, -56N and Hg: -0.87 to -0.69), and elbow flexion (p: 0.004, -85N and Hg: -1.44 to -1.30), see also Appendix table 2.

3.7. Movement performance

There was a significant support effect for task duration, efficiency, and jerk, and a significant group * support interaction effect for jerk, with longer task duration with support, reduced efficiency with support and reduced jerk with support in both groups (Appendix figure 14).

Additionally, there were significant task effects (task duration and jerk), significant group * task interaction effects (task duration and efficiency), and significant support * task interaction effects (efficiency and jerk) found.

4. Discussion

4.1. Muscle coordination consistency and similarity

In this study, we investigated muscle coordination in persons with FSHD and healthy controls when performing ADL, without and with the use of a dynamic arm support device. Our first hypothesis was partially accepted, as without support muscle coordination was less consistent in FSHDs than controls for the
4.2. Muscular weakness in persons with FSHD

This is the first study to examine muscle coordination synergies in persons with FSHD during ADL. Our findings with regards to synergy weights during unsupported tasks are consistent with previous results in single joint arm elevation movements [13], revealing that muscle coordination in persons with FSHD remains heterogeneous during the ADL tasks used in this study. The nature of the unsupported task, that being whether it was close or away from the body, appears to influence the synergies’ consistency of both the first (task effect) and the second ranked synergies (task effect and support * task interaction effect) within each group.

A clear-cut categorization of the synergies based on muscle function is not straightforward, but we made the following observations in control participants. During unsupported tasks, the first ranked synergy mostly involved the muscles responsible for elevation, rotation of the scapula, and arm adduction, while the second ranked synergy mostly involved those muscles responsible for scapula external rotation and arm abduction. Observation of the synergy weights in control participants reveals that, in unsupported tasks that are closer to the body, MS1 was characterized by a prominent involvement of the trapezius muscle, in tasks far away from the body, the deltoid was also involved. In MS2, the trapezius, serratus, and latissimus were largely involved during unsupported tasks that were closer to the body (cup and spoon to mouth), with the deltoid becoming additionally involved in unsupported tasks away from the body (reaching). In the push and pull task, which consisted of a reach and retrieval phase, the trapezius was not involved. The involvement of the trapezius and serratus during far away from the body tasks, where arm elevation was necessary to reach the target, is consistent with the functional anatomy. Both these muscles are in fact necessary to accomplish scapular lateral rotation [39].

In FSHD participants, a higher level of muscle co-contraction compared to controls was present for all muscle weights in both
synergies during all unsupported tasks. This higher level of co-contractation in the FSHD weights was also accompanied by a higher variation in neural activation, as shown by the coefficients. Furthermore, higher maximum muscle activity was found for the biceps, deltoid, pectoralis, and latissimus dorsi compared to controls, which is in line with previous literature during unsupported tasks [7]. Despite these heterogeneous neuromuscular activations and the lower muscle strengths found in FSHD, the movement performance indicators could not be shown to differ between the two groups. These findings indirectly highlight the existence of compensatory movement strategies in persons with FSHD that aid task completion but also lead to a greater muscle effort than controls.

4.3. The effects of dynamic arm support

The effects of a dynamic arm support on motor capacity, i.e. what a person can do in controlled settings, are reported for the first time in persons with FSHD. Knowledge of motor capacity and capability are important to assess how an arm support is used and ultimately to better understand the reasons a person may discontinue its use [10,40].

When using the arm support, both groups displayed a reduction in maximum muscle activity of the biceps, deltoid, triceps, serratus, and latissimus. Yet, a more generalized co-activation was apparent in all muscles in the FSHD group. Internal consistency in this group was not significantly affected despite general alterations in muscle activity. The increased synergy similarity between the control and FSHD groups when using the support illustrates that the FSHD group did alter their synergies when assisted by the support device. The internal consistency, however, was lower in the FSHD than the control group and the group differences grew larger with the use of an arm support. These novel findings indicate that muscle coordination in persons with FSHD remains heterogeneous, which is likely the result of the individual-specific deficits in muscle strength.

Although the dynamic arm support facilitated arm elevation, the device also affected movement performance by restricting range of motion and increased movement duration in both groups. These findings are consistent with the existing literature in healthy adults and in stroke patients [15,16,19]. Future research should investigate the movement dynamics with the use of arm support devices, as it would be useful in persons with FSHD, to better understand how joint forces, moments, and powers are affected by the devices, in particular whether eccentric-concentric contractions are employed in response to the gravity compensation. Knowledge of these adaptations may have implications for a more efficient design of the arm support device and for the prevention of long-term neck-shoulder complaints, which are reported by more than 90% of adults with FSHD [6].

4.4. Limitations

The tasks were always completed first without device and then with device, which could have affected the results due to a higher chance of fatigue in the latter condition. However, resting periods, a randomization of the tasks, and repetitions were incorporated in both conditions to minimize fatigue and ensure that participants could complete the study.

The limited number of three repetitions per task, used in the current study, could have reduced the internal consistency in all cases, but would not have affected the number of synergies extracted [41]. Moreover, the number of repetitions was experienced as very demanding by some FSHD participants and more repetitions would likely have resulted in discontinuation of the study.

The dynamic arm support, Gowing, imposed mechanical constraints that affected the performance of both groups. The elbow brace was experienced as a slight inconvenience during tasks away from the body, but did not hinder the participants in task execution. Participants could have compensated for the inconvenience, resulting in a more variable execution, but there were no indications noted in the outcome parameters.

4.5. Considerations for future research

Scapular kinematics and activity of deeper-layered muscles, such as the rhomboids, teres, and supra- and infraspinatus, should be considered in future research to better understand the effect of arm supports on scapular mobility and glenohumeral stabilization.

Future research should also consider the long-term effects of using a dynamic arm support device in a home environment to uncover potential benefits and disadvantages associated with regular, home use. The positive effects on motor capacity from the current study might also be reflected in long-term benefits in motor performance by experienced dynamic arm support users during repetitive tasks [9]. Negative effects, such as discomfort or inconvenience to perform certain ADL, may add to the evidence for discontinued use of the support device.

Conclusion

We found that muscle coordination is altered and less consistent in FSHDs compared with healthy controls. An arm support alleviated muscle efforts and affected muscle coordination in both populations by facilitating arm elevation. Consequently, the populations became more similar, yet, the internal consistency of FSHDs remained unaffected and lower than that of healthy controls. This is likely the result of the individual-specific deficits of muscle weakness and respective development of compensatory strategies for dealing with the compensation of gravity and movement constraints. The biomechanical consequences of using an arm support should be further investigated in people with FSHD on deeper-layered shoulder muscles and to evaluate potential long-term benefits and disadvantages.

Declaration of Competing Interest

All authors indicate that there are no (financial) conflicts of interest. All authors take responsibility for the contents of the manuscript and satisfy the requirements for authorship.

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Supplementary materials

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