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Reachable workspace reflects dynamometer-measured upper extremity strength in FSHD

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Abstract

Introduction—It is not known if a reduction in reachable workspace closely reflects loss of upper extremity strength in facioscapulohumeral muscular dystrophy (FSHD). This study aims to determine the relationship between reachable workspace and quantitative upper extremity strength measures.

Methods—Maximal voluntary isometric contraction (MVIC) testing of bilateral elbow flexion and shoulder abduction by hand-held dynamometry was performed on 26 FSHD and 27 control subjects. Additionally, Kinect sensor-based 3D reachable workspace relative surface areas (RSA) were obtained. Loading (500-gram weight) effects on reachable workspace were also evaluated.

Results—Quantitative upper extremity strength (MVIC of elbow flexion and shoulder abduction) correlated with Kinect-acquired reachable workspace RSA ($R=0.477$ for FSHD, $P=0.0003$; $R=0.675$ for the combined study cohort, $P<0.0001$). Progressive reduction in RSA reflected worsening MVIC measures. Loading impacted the moderately-weak individuals the most with additional reductions in RSA.

Discussion—Reachable workspace outcome measure is reflective of upper extremity strength impairment in FSHD.

MESH terms

Reachable workspace; upper extremity; dynamometry; FSHD; Kinect

Introduction

Facioscapulohumeral muscular dystrophy (FSHD) is 1 of the most common muscular dystrophies with approximate prevalence of 1:15,000–1:20,000.^{1,2,3} It is an autosomal dominantly inherited muscular dystrophy caused by loss of sub-telomeric *D4Z4* macrosatellite repeats that contain copies of *DUX4* homeodomain retrogene.^{4,5,6} Although

facial and lower extremity function can be affected in FSHD, the stereotypical and progressive loss of muscle strength in the shoulder girdle leads to its most notable and significant functional impairment in the upper extremities.⁷

Monitoring upper extremity strength decline and functional loss in FSHD can be achieved by functional scales (FSHD clinical evaluation scale), manual muscle testing (MMT),⁹ or quantitative muscle testing (QMT).¹⁰ As a direct quantitative measure of muscle strength, maximal voluntary isometric contraction (MVIC) testing in FSHD has been performed via either formal QMT^{9,10} or through the use of a hand-held dynamometer.¹¹

Recently, there has been significant progress in understanding the underlying pathophysiology of FSHD and identification of promising therapeutic targets.¹² However, these advances have also highlighted a critical need to develop innovative clinical outcome tools that can accurately monitor disease progression and effectively evaluate the efficacy of these promising strategies.

Taking advantage of currently available unobtrusive markerless sensor technology, a novel upper extremity outcome measure based on Kinect sensor-acquired 3D reachable workspace was recently developed by the study investigators^{13,14} and applied in FSHD.¹⁵ In the previous study, the novel reachable workspace outcome measure demonstrated its validity against the upper extremity-specific components of the FSHD evaluation scale (subscales II: scapular girdle and III: upper limb). In addition, it demonstrated excellent discriminative ability to differentiate not only FSHD individuals from healthy controls, but also the capability to differentiate among FSHD individuals with minimal, moderate, and severe impairments of upper extremity function.¹⁵

In this study, we examine if the newly-developed Kinect-acquired reachable workspace outcome measure also directly reflects upper extremity strength impairment found across a range of disease severity in FSHD. We examine whether the reachable workspace outcome measure (relative surface area, RSA) correlates well with a quantitative upper extremity strength measure (MVIC) and can serve as a novel surrogate marker of disease progression in FSHD. Additionally, we assess the sensitivity of the developed outcome measure system to detect change by evaluating the differential effects of a simple loading protocol (500-gram wrist weight) on the reachable workspace of individuals with FSHD.

MATERIALS AND METHODS

Study Participants

A total of 26 subjects with FSHD (12 women and 14 men) and 27 healthy controls (12 women and 15 men) participated in the study. All study participants with FSHD had a confirmed diagnosis by DNA testing showing a reduced number of sub-telomeric *D4Z4* macrosatellite repeat units. Healthy controls without any neuromuscular or musculoskeletal disorders were recruited from the community. For the study, an age-, gender-, height-, and weight-matched control cohort was assembled. This was to allow for reference hand-held dynamometry strength measures from healthy controls to be compared to the FSHD cohort data. Demographic and anthropometric information (age, gender, hand-dominance, height,

and weight) were collected from each subject. The study protocol was approved by the University Institutional Review Board (IRB) for human protection and privacy for research. All participants provided informed written consent prior to participation in the study.

Hand-held dynamometry

MVIC was measured with a MicroFet2 hand-held dynamometer (Hogan Health Industries, Inc. West Jordan, USA). Strength was measured using an isometric “make” technique from a previously described protocol¹⁶ modified to accommodate neuromuscular subjects. Briefly, the study subjects were asked to build up force to maximum over the course of 3–4 seconds. Isometric muscle force (N, Newton) of elbow flexion and shoulder abduction were measured. Three consecutive measurements were performed with adequate rest intervals between contractions. For elbow flexion, subjects were seated with the arm alongside the body, elbow flexed at 90°, and forearm supinated. The dynamometer was placed over the volar aspect of the distal forearm just proximal to the wrist crease. For shoulder abduction, subjects were seated with the shoulder abducted at 90° and elbow flexed at 90° with palm facing down. The dynamometer was placed just proximal to the lateral epicondyle. For each subject, an average raw muscle force (N) was calculated from the 3 trials for both the dominant and non-dominant sides. Each subject’s mean raw quantitative strength data (MVIC) were then normalized to predicted reference values for healthy controls (% predicted) based on age, gender, height, weight, and hand-dominance using the regression equations for calculating reference values.¹⁷ To obtain the percent predicted combined elbow flexion and shoulder abduction strength, the following formula was used: $[\text{Raw Elbow Flexion (N)} + \text{Raw Shoulder Abduction (N)}] / [\text{Predicted Normal Elbow Flexion (N)} + \text{Predicted Normal Shoulder Abduction (N)}] \times 100$.

Upper Extremity Reachable Workspace Protocol

The sensor system setup and arm movement detection followed the already published protocol.^{14,15} The subjects were seated in front of the Microsoft Kinect sensor (Microsoft Corp., Redmond, WA, USA) and performed a standardized upper extremity movement protocol under the supervision of a study clinical evaluator. Briefly, the upper extremity movements consisted of lifting the arm from the resting position to above the head while keeping the elbow extended, performing the same movement in vertical planes at around 0, 45, 90, and 135 degrees, and horizontal sweeps at the level of the umbilicus and shoulder. After the non-weighted reachable workspace data was collected, all subjects also underwent a loading protocol with a wrist weight (500-gram). For test-retest reliability/reproducibility assessments, the subjects returned several hours later to undergo a repeat data collection on the same day.

3D Reachable Workspace Surface Envelope Analysis (relative surface area, RSA)

The Kinect-acquired 3D hand trajectory was transformed into a body-centric coordinate system, and each individual’s reachable workspace surface envelope was reconstructed in a graphical output using methods described previously.¹⁴ Each arm’s reachable workspace envelope was further divided into 4 quadrants with the shoulder joint serving as the origin: above and below the shoulder joint, ipsilateral and contralateral side relative to the side being examined (Figure 1). The absolute total and quadrant reachable workspace surface

envelope areas (m^2) were calculated and normalized by each individual's arm length and unit hemi-sphere to obtain the relative surface area (RSA) following a method described previously.¹⁵

Statistical Analyses

Statistical analyses used SAS 9.4 software. Data were checked for normality and analyzed parametrically. Pearson correlations were used to determine associations of parametric data, and to examine test-retest reliability. Student *t*-tests were used to assess differences between 2 groups. An ANOVA was used to assess differences between multiple groups, and a *post-hoc* Tukey analysis was used to determine sub-group differences. For all statistical analyses, a minimum *P*-value of 0.05 was accepted as the level of statistical significance.

RESULTS

Study participants

Twenty-six individuals with FSHD and 27 healthy controls participated in the study. The FSHD and healthy control cohorts were well-matched for age, height and weight; a summary of the study participant characteristics is shown in Table 1. Of 26 FSHD subjects, 3 were left-handed while 1 of 27 controls was left-handed.

Test-retest reliability of Kinect-acquired reachable workspace in FSHD

The test-retest reliability of the automated Kinect-acquired 3D reachable workspace in a group of healthy controls was reported previously.¹⁴ Test-retest reliability of the Kinect-acquired reachable workspace in FSHD was examined in this study with a subset of upper extremities from the FSHD cohort ($n=8$, mean age 37.3 ± 24.8 years, height 164.1 ± 15.3 cm, weight 60.04 kg). These assessments were completed with the Kinect upper extremity movement protocol at the beginning and end of the study visit, but not all participants completed both assessments. The Pearson correlation coefficient for test-retest reliability was $R=0.952$ ($P<0.0001$) for FSHD.

Upper Extremity MVIC measures by hand-held dynamometry

The hand-held dynamometry data of the 26 FSHD subjects (raw and %-predicted) is shown in Table 2 (the control group data are not shown in a detailed table format, but are represented in Supplementary Table S1, available online). The raw MVIC data for each subject by handedness, as well as for both elbow flexion and shoulder abduction, are shown along with %-predicted MVIC (as compared to predicted reference values by age, sex, height, weight, and hand-dominance) in Supplementary Table S1. Also, shown is the combined elbow flexion and shoulder abduction %-predicted MVIC of each FSHD subject by hand-dominance. Graphical illustrations depicting the distribution of the normalized strengths (%-predicted MVIC) of both the FSHD and control cohorts (combined dominant/non-dominant) are shown in Figure 2.

Correlation between reachable workspace (RSA) and hand-held dynamometry (%-predicted MVIC)

Correlations between normalized %-predicted MVIC (elbow flexion, shoulder abduction, and combined) and Kinect-acquired total reachable workspace RSA in the FSHD cohort and the entire study cohort (FSHD + Controls) are shown in Figure 3. For the FSHD cohort, moderate correlation with reachable workspace was observed for elbow flexion ($R=0.528$, $P<0.0001$), while no significant correlation was observed for shoulder abduction quantitative strength ($R=0.094$, $P=0.508$). Similarly moderate correlation was noted for combined elbow flexion and shoulder abduction %-predicted MVIC and the reachable workspace RSA ($R=0.477$, $P=0.0003$). Looking at the entire study cohort (combined FSHD and control subjects together), all upper extremity quantitative strength measures including elbow flexion and shoulder abduction separately, as well as the combined elbow and shoulder strength measures demonstrated significant correlation with the total reachable workspace RSA (elbow flexion: $R=0.664$; shoulder abduction: $R=0.479$; combined elbow + shoulder: $R=0.675$, all with $P<0.0001$). *Post hoc* Student *t*-test analyses revealed no significant differences between men and women in total RSA in the FSHD ($P=0.845$) or control cohorts ($P=0.497$).

Decreased upper extremity strength is reflected by progressive reduction in reachable workspace

Visualized through a reachable workspace plot (Figure 4), the total and quadrant reachable workspace RSAs for the FSHD cohort are stratified to represent degrees of weakness (mild, moderate, severe, and extreme weakness) by their upper extremity MVIC strength based on 4 categories: $>80\%$, 50 to 80%, 20 to $<50\%$, and $<20\%$ -predicted. As can be observed, the overall trend shows a strong direct relationship between reachable workspace RSA and the elbow flexion MVIC strength measure (Fig. 4A), while less evident overall trend can be appreciated with shoulder abduction (Fig. 4B). The most clear relationship between progressive weakness of the upper extremity and reduction in reachable workspace can be observed in Figure 4C, where combined progressive weakness in elbow flexion and shoulder abduction results in gradually decreasing reachable workspace RSA (the weaker individuals are, based on hand-held dynamometry of elbow flexion and shoulder abduction, the smaller their reachable workspace RSA tends to be).

Representative illustrations of progressive reduction in reachable workspace observed with decreasing upper extremity MVIC measures (combined %-predicted strength of elbow flexion and shoulder abduction) are shown in Figure 5. The paired figures (left and right) also show the effect of the loading condition (500-gram wrist weight) on the reachable workspace RSA of individuals with varying degrees of upper extremity weakness.

Sensitivity of the reachable workspace and evaluation with loading protocol (wrist-weights)

The sensitivity of reachable workspace outcome to detect change in upper extremity function was assessed by examining the effects of different loading conditions (non-weighted vs. 500-gram wrist weight) on reachable workspace (Fig. 6). A change in total reachable workspace RSA with addition of a 500-gram wrist weight is shown in Figure 6A.

Minimal change is observed in healthy controls and in individuals with FSHD who have relatively preserved upper extremity strength (>80%). For individuals with FSHD who are extremely weak (<20%), the reachable workspace is already considerably reduced to begin with, and little additional change is noted with addition of the 500-gram wrist weight. The greatest change in reachable workspace produced by addition of 500-gram wrist weight is noted for individuals who are moderately-weak at 50–80% predicted MVIC for the combined upper extremity strength. A closer examination of the amount of RSA changes in both quadrant and total reachable workspace due to the 500-gram wrist weight reveals that in individuals with moderate upper extremity weakness (50 to 80%), there is approximately a 6% reduction in total reachable workspace RSA that is significantly different from the controls (Fig. 6B). Reachable workspace reductions in quadrants 1 and 3 (upper contralateral and ipsilateral quadrants respectively), and 4 (lower ipsilateral quadrant) accounted for the majority of the total reachable workspace reduction due to loading.

DISCUSSION

Recent developments of promising therapeutic strategies for FSHD have identified an important need for improved understanding of disease progression to better inform the design of future clinical trials.¹⁸ Moreover, it has highlighted a critical need to identify outcome measures that can accurately monitor disease progression and detect changes with adequate sensitivity to evaluate the efficacy of promising therapeutic interventions. Traditionally, MMT and QMT (MVIC) outcome measures have been utilized to evaluate strength in FSHD, and both have demonstrated good validity and high reliability as outcome measures.^{9,10} However, a recent review of a number of natural history studies and clinical trials that have utilized the MMT and MVIC methods in FSHD found that these measures may need re-evaluation as outcome measures to detect significant changes over the typical time course of a clinical study.¹⁸

In part responding to the need to develop novel outcome measures, the FSHD evaluation scale and clinical score was developed specifically to provide an overall assessment of disease severity by measuring the functionality of the 6 major muscle groups involved in the disease (subscales I–VI).⁸ In particular, the subscales II (scapular girdle) and III (upper limb) focus specifically on clinical evaluation of upper extremity function. However, the ordinal nature of these scores and relative lack of granularity to detect subtle changes in upper extremity function over the relatively short time period typical of a clinical study may pose problems for a slowly progressive disease such as FSHD.

With these factors to consider and being mindful of various other practical qualities in development of outcome measures such as ease-of-use, cost-effectiveness, and scalability, we have developed a low-cost single sensor-based 3D reachable workspace assessment system for upper extremity function.^{13,14,15,19,20} A proof-of-concept as well as a series of feasibility studies have led to clinical application of the system in FSHD, where its stereotypical shoulder girdle strength impairment may be an ideal model to examine the reachable workspace outcome measure.¹⁵ In a previously published study, the Kinect-based reachable workspace assessment system demonstrated its concurrent validity against the

FSHD functional scale (the upper extremity subscales II and III of the FSHD evaluation scale).¹⁵

In this study, the main aim was to determine whether the reachable workspace can also directly reflect upper extremity strength loss in FSHD. This was examined by comparing Kinect-acquired reachable workspace RSA against quantitative measures of upper extremity strength (hand-held dynamometer MVIC of elbow flexion and shoulder abduction).

Prior to comparison however, we first assessed the test-retest reliability of the Kinect reachable workspace outcome measure in FSHD and found it to be excellent ($R=0.952$, $P<0.0001$). Next, the quantitative strength measures of elbow flexion and shoulder abduction by hand-held dynamometer for the study FSHD cohort and controls were examined. The normalized quantitative strength measure (%-predicted MVIC) showed clear separation between the FSHD and control cohorts with differential distribution of strength impairment in the FSHD cohort compared to the healthy controls (Fig. 2). The healthy control strength values centered around the expected 100%-predicted reference values with a normal distribution, and this provided further assurance of the internal validity of the study protocol involving the quantitative strength measures. We next proceeded to assess a more detailed relationship between the Kinect-acquired reachable workspace and the upper extremity MVIC strength measures.

There was a strong correlation between quantitative upper extremity strength and the reachable workspace outcome measure (Fig. 3). Although elbow flexion correlated with the reachable workspace better than shoulder abduction, the combined elbow flexion and shoulder abduction quantitative strength measure correlated the best with the reachable workspace. This can also be visualized graphically in Figure 5C, where the combined upper extremity strength measure stratified by the degree of weakness (mild, moderate, severe, and extreme weakness) demonstrates a clear sequential relationship with the reachable workspace RSA; a progressive weakness in combined upper extremity strength (that includes both elbow flexion and shoulder abduction) demonstrated gradual reduction in reachable workspace. The study's findings also suggest that the reachable workspace outcome measure may have the advantage of incorporating both the elbow and shoulder strength impairments in 1 surrogate functional outcome measure. Further studies will need to be conducted to examine not only the relationship between reachable workspace with functional scales and strength measures (as shown in this and the previously published study¹⁵), but also with person-reported upper extremity function itself.

The sensitivity of the reachable workspace outcome measure to further differentiate between individuals with subtle differences in strength by the loading protocol is also shown here. Although the study only examined the effect of 1 weight (500-gram), the findings show the potential capability of the reachable workspace outcome measure to further differentiate individuals with moderate weakness (50–80%) through the use of a loading protocol. This may be an important quality of the outcome measure, as this moderately-weak group of individuals may be the most likely target to show efficacy in therapeutic intervention trials. Although the small sample size precludes an in-depth and detailed subgroup statistical analysis, the potential to use a loading protocol with wrist-weights as a simple method to

improve the sensitivity to detect strength loss (or strength improvement in response to a therapeutic intervention) provides additional testing protocol option/flexibility for clinical studies in FSHD and is a promising area for future investigation.

Limitations of the study include the relatively small sample size and quantitative upper extremity strength measures that were limited to elbow flexion and shoulder abduction. Inclusion of other muscle groups for a more detailed strength analysis (e.g. elbow extensors and shoulder forward flexors) would be interesting, and potentially may help to increase the correlation seen with the reachable workspace. We noted that shoulder abduction strength by itself had a poor correlation with reachable workspace. This may be because although shoulder abduction likely plays a role in affecting reachable workspace (as demonstrated in this study), it may not be the prime contributor to significantly impact the ‘frontal’ hemisphere reachable workspace measure. In fact, shoulder forward flexion may have a closer association with reachable workspace. Further studies with a larger sample size to examine these issues, as well as a longitudinal natural history study examining the sensitivity of the outcome measure to track disease progression would be helpful. In this respect, however, a distinct advantage of the newly-developed sensor-based and automated reachable workspace assessment system is its scalability, ease-of-use, low-cost, and minimal equipment requirements to facilitate such large-scale, multi-site, and international investigations.

Advances in sensor technology and digital health tools are infusing novel ideas into clinical outcomes development that traditionally have relied on time- and effort-intensive methods. Development of an innovative surrogate outcome measure that evaluates upper extremity functional performance such as reachable workspace, rather than the traditional measurements of static muscle strength, may be more sensitive to changes in muscle function and represent a promising area for future investigations.

CONCLUSION

This study shows that the Kinect sensor-acquired reachable workspace outcome measure reflects upper extremity strength impairment in FSHD. The demonstrated validity, reliability, and greater sensitivity to detect changes through use of a loading protocol, in combination with scalability of the system, makes the Kinect-based reachable workspace assessment and outcome measure system a promising new clinical tool in the FSHD field. Continued development of innovative outcome measures will not only improve our understanding of disease progression in natural history studies, but will also facilitate the overall therapeutic discovery process towards finding an effective treatment.

Supplementary Material

Refer to Web version on PubMed Central for supplementary material.

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ABBREVIATIONS

3D	Three-dimensional
ANOVA	Analysis of variance
FSHD	Facioscapulohumeral muscular dystrophy
MMT	Manual muscle test
MVIC	Maximal voluntary isometric contraction
QMT	Quantitative muscle test
RSA	Relative surface area

REFERENCES

1. Tawil R. Facioscapulohumeral muscular dystrophy. *Neurotherapeutics*. 2008 Oct; 5(4):601–606. [PubMed: 19019312]
2. Flanigan KM, Coffeen CM, Sexton L, Stauffer D, Brunner S, Leppert MF. Genetic characterization of a large, historically significant Utah kindred with facioscapulohumeral dystrophy. *Neuromuscul Disord*. 2001; 11(6–7):525–529. [PubMed: 11525880]
3. Mostacciolo ML, Pastorello E, Vazza G, Miorin M, Angelini C, Tomelleri G, et al. Facioscapulohumeral muscular dystrophy: epidemiological and molecular study in a north-east Italian population sample. *Clin Genet*. 2009; 75(6):550–555. [PubMed: 19320656]
4. van Deutekom JC, Wijmenga C, van Tienhoven EA, et al. FSHD associated DNA rearrangements are due to deletions of integral copies of a 3.2 kb tandemly repeated unit. *Hum Mol Genet*. 1993; 2:2037–2042. [PubMed: 8111371]
5. Wijmenga C, Hewitt JE, Sandkuijl LA, et al. Chromosome 4q DNA rearrangements associated with facioscapulohumeral muscular dystrophy. *Nat Genet*. 1992; 2:26–30. [PubMed: 1363881]
6. Van der Maarel SM, Tawil R, Tapscott SJ. Facioscapulohumeral muscular dystrophy and DUX4: breaking the silence. *Trends Mol Med*. 2011; 17(5):252–258. [PubMed: 21288772]
7. Kilmer DD, Abresch RT, McCrory MA, Carter GT, Fowler WM Jr, Johnson ER, McDonald CM. Profiles of neuromuscular diseases. Facioscapulohumeral muscular dystrophy. *Am J Phys Med Rehabil*. 1995 Sep-Oct; 74(5 Suppl):S131–S139. [PubMed: 7576420]
8. Lamperti C, Fabbri G, Vercelli L, D'Amico R, Frusciante R, Bonifazi E, et al. A standardized clinical evaluation of patients affected by facioscapulohumeral muscular dystrophy: The FSHD clinical score. *Muscle Nerve*. 2010; 42:213–217. [PubMed: 20544930]
9. Tawil R, McDermott MP, Mendell JR, Kissel J, Griggs RC. Facioscapulohumeral muscular dystrophy (FSHD): design of natural history study and results of baseline testing. FSH-DY Group. *Neurology*. 1994 Mar; 44(3 Pt 1):442–446. [PubMed: 8145913]
10. Personius KE, Pandya S, King WM, Tawil R, McDermott MP. Facioscapulohumeral dystrophy natural history study: standardization of testing procedures and reliability of measurements. The FSH DY Group. *Phys Ther*. 1994 Mar; 74(3):253–263. [PubMed: 8115459]
11. Brouwer OF, Padberg GW, van der Ploeg RJ, Ruys CJ, Brand R. The influence of handedness on the distribution of muscular weakness of the arm in facioscapulohumeral muscular dystrophy. *Brain*. 1992 Oct; 115(Pt 5):1587–1598. [PubMed: 1422805]
12. Statland JM, Tawil R. Facioscapulohumeral muscular dystrophy: molecular pathological advances and future directions. *Curr Opin Neurol*. 2011 Oct; 24(5):423–428. [PubMed: 21734574]

13. Kurillo G, Han JJ, Obdržálek S, Yan P, Abresch RT, Nicorici A, Bajcsy R. Upper extremity reachable workspace evaluation with Kinect. *Stud Health Technol Inform*. 2013; 184:247–253. [PubMed: 23400165]
14. Kurillo G, Chen A, Bajcsy R, Han JJ. Evaluation of upper extremity reachable workspace using Kinect camera. *Technol Health Care*. 2013; 21(6):641–656. [PubMed: 24284552]
15. Han JJ, Kurillo G, Abresch RT, de Bie E, Nicorici A, Bajcsy R. Reachable Workspace in Facioscapulohumeral muscular dystrophy (FSHD) by Kinect. *Muscle Nerve*. 2014 May 15. [Epub ahead of print].
16. Bohannon RW. Reference values for extremity muscle strength obtained by hand-held dynamometry from adults aged 20 to 79 years. *Arch Phys Med Rehabil*. 1997 Jan; 78(1):26–32. [PubMed: 9014953]
17. Douma RK, Soer R, Krijnen WP, Reneman M, van der Schans CP. Reference values for isometric muscle force among workers for the Netherlands: a comparison of reference values. *BMC Sports Sci Med Rehabil*. 2014 Feb 25.6(1):10. [PubMed: 24568140]
18. Statland JM, McDermott MP, Heatwole C, Martens WB, Pandya S, van der Kooi EL, Kissel JT, Wagner KR, Tawil R. Reevaluating measures of disease progression in facioscapulohumeral muscular dystrophy. *Neuromuscul Disord*. 2013 Apr; 23(4):306–312. [PubMed: 23406877]
19. Kurillo G, Han JJ, Abresch RT, Nicorici A, Yan P, Bajcsy R. Development and application of stereo camera-based upper extremity workspace evaluation in patients with neuromuscular diseases. *PLoS One*. 2012; 7(9)
20. Han JJ, Kurillo G, Abresch RT, Nicorici A, Bajcsy R. Validity, Reliability, and Sensitivity of a 3D Vision Sensor-based Upper Extremity Reachable Workspace Evaluation in Neuromuscular Diseases. *PLoS Curr*. 2013 Dec 12.:5.

Figure 1. Graphical visualization of Kinect-based 3D reachable workspace assessment system
An example graphical output with quadrant designation is shown (right shoulder perspective is shown).

Figure 2. Distribution of quantitative upper extremity strength measures for the study cohorts
 The hand-held dynamometry upper extremity strength measures (%-predicted MVIC) for both the FSHD and control groups are shown (combined data for men and women, dominant and non-dominant sides). (A) Elbow flexion (FSHD: 57.21 ± 39.22 , $n=52$ vs. Control: 96.58 ± 18.25 , $n=52$; $P < 0.0001$). (B) Shoulder abduction (FSHD: 52.57 ± 24.96 , $n=52$ vs. Control: 91.57 ± 20.69 , $n=54$; $P = 0.508$). (C) Combined elbow flexion and shoulder abduction (FSHD: 55.40 ± 27.94 , $n=52$ vs. Control: 94.09 ± 15.68 , $n=52$; $P < 0.0001$).

Figure 3. Correlation between hand-held dynamometry (%-predicted MVIC) and the Kinect reachable workspace RSA

(A) Elbow flexion – for the FSHD cohort: $R=0.528$, $P<0.0001$; for the entire study cohort including FSHD + Control: $R=0.664$, $P<0.0001$. (B) Shoulder abduction – for FSHD: $R=0.094$, $P=0.508$; the entire group: $R=0.479$, $P<0.0001$. (C) Combined elbow flexion + shoulder abduction – for FSHD: $R=0.477$, $P=0.0003$; the entire group: $R=0.675$, $P<0.0001$. Solid line = regression line for the FSHD cohort; Dotted line = regression line for the entire FSHD + Control subjects.

Figure 4. Reachable workspace plot showing total and quadrant RSAs of control and FSHD cohorts stratified by quantitative strength measures
 (A) Elbow flexion, (B) Shoulder abduction, and (C) Combined elbow flexion and shoulder abduction (sample sizes: >80%, n=10; 50 to 80%, n=21; 20 to <50%, n=17; and <20%, n=4).

Figure 5. Paired figures showing non-loading (left) and 500-gram wrist weight effects (right) on RSAs of representative individuals

The figures demonstrate the reachable workspace of individuals from the following groups: (A) Control, and FSHD with %-predicted combined elbow flexion and shoulder abduction MVIC strength categories of (B) >80%, (C) 50 to 80%, (D) 20 to <50%, and (E) <20%.

Figure 6. Changes in reachable workspace with loading

(A) A line graph showing changes in mean total RSA with loading (500-g wrist weight), in controls and the FSHD cohort categorized by %-predicted upper extremity strength, and (B) quadrant and total RSA with loading ($\Delta \text{RSA} = \text{RSA with 500g load} - \text{RSA with no load}$), in controls and the FSHD cohort categorized by %-predicted upper extremity strength (*significantly different from controls, $P < 0.05$).

Table 1

Study participant characteristics

	Gender (M/W)	FSHD Women: n=12 Men: n=14	Control Women: n=12 Men: n=15	P-value
Age (yrs)	Women	45.9±24.4	40.5±22.5	0.433
	Men	51.1±18.0	45.3±15.0	0.678
Height (cm)	Women	106.2±7.3	163.0±11.9	0.326
	Men	178.2±6.8	178.8±6.5	0.722
Weight (kg)	Women	66.4±23.7	65.6±17.7	0.890
	Men	87.9±13.1	90.7±14.7	0.446