AI-based wearable device for tracking shoulder joint kinematics and muscle activity in subjects with FSHD disease

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INTRODUCTION: Facioscapulohumeral muscular dystrophy (FSHD) results from epigenetic derepression in skeletal muscles of the double homeobox 4 (DUX4) gene located at chromosome 4q35. FSHD is among the most common muscular dystrophies, affecting between 1 in 8000 and 1 in 20000 individuals in different populations, with a variable age of onset, severity, and progression. There is no definitive cure for FSHD, but exciting pre-clinical research studies have identified an increasing number of candidate therapeutics targeting aberrant DUX4 expression that are now ready to be evaluated rigorously in clinical trials. To the extent that disease progression in FSHD as captured by global outcome measures is slow, it may be difficult to obtain informative results in a trial without extending the trial duration or focusing upon more specific aspects of muscle function[1]. As the field is advancing for new trials in FSHD, there is an acute need for valid, reliable, and quantitative clinical metrics to assess FSHD and monitor disease progression, especially those that reflect the functions most relevant to daily life for affected patients. Not only are these clinical outcome measurements useful in clinical trials, but they can also provide valuable information on disease severity or progression in natural history studies. Neuromuscular disease clinical trials frequently have focused on leg function, ambulation, and mobility (e.g., timed walk test) as well as lower extremity outcome measures (e.g., time to climb 4 stairs or the 10 meter/walk run test). However, assessment of shoulder girdle and upper extremity function is critical to emphasize in FSHD clinical studies because it is tied closely to patients' basic self-care activities of daily living, quality of life, and independence. Although these functions have been evaluated by manual and quantitative muscle testing with good validity and high reliability innovative new methods may be needed to identify significant changes over time for FSHD studies. In addition to challenges of in-person evaluation and clinical variability, assessment of strength and range of motion (ROM) may also be biased by the patients' cooperation or effort during the examination, ceiling effects, floor effects, and the investigators' experience.

METHODS: We propose here an innovative clinical outcome assessment using an A.I.-based wearable device for tracking shoulder joint kinematics and muscle activity in FSHD subjects compared to age-, gender-, and height-matched healthy controls. The device is a mobile app controlled wireless wearable adjustable upper arm/shoulder brace that can monitor and measure joint range of motion and muscle activity, transmit these data wirelessly to a mobile app, then along to cloud storage for data analysis. Our proposed technology will provide quantitative and reliable measurement of shoulder ROM and muscle activity with remote data-sharing capacity that can be used longitudinally in both FSHD clinical trials and natural history studies. It can also decrease medical costs while increasing patient engagement through goal-oriented disease progression feedback. This innovative technology will link clinically relevant pathologies to the mechanics and functions of individual muscles affected by FSHD to inform regarding the phenotypic spectrum of shoulder girdle muscle involvement in FSHD. In addition to developing a novel outcome measure to be utilized in natural history studies and clinical trials, this approach shows potential for remote telemedicine delivery of customized rehabilitation services.

Figure.1. Proposed conceptual intelligent wearable device

Our device was utilized in a pilot study of healthy subjects and patients with FSHD. Subjects initially stand upright in the neutral position for 5 to 10 s, with their arms relaxed by their sides (palms inward, facing the body). Sensor data will be recorded for 10 s to establish sensor baseline orientation and noise. Each subject will perform four active and passive movements: shoulder abduction, elbow flexion, elbow extension, and shoulder external rotation. In each performance, subjects will have a 3-minute break to avoid any muscle fatigue and tiredness. To assess the reliability, three repeated trials is performed on the same day for each side and also in two different sessions after a baseline visit at 1 and 3 months. Calculated and measured variables are summarized in Table.1. A one-way repeated measure of analysis of variance (ANOVA) is used to determine the reliability during each activity. Individual error scores of zero indicate reliability. We also examine the interclass correlation coefficient (ICC) of each of the three trials and make Bland-Altman plots for each session as well as for the three different sessions (baseline, month 1, and month 3).

Table 1. Summary of Outcome Variables

Our Device Outcomes	Other FSHD outcomes in our study
ROM (degree) for each performance (*i.e.,* shoulder abduction, elbow flexion, elbow extension, and shoulder external rotation) during the going phase (0-180°)	Brooke upper extremity functional scale
ROM (degree) for each performance during returning phase (180-0°)	Medical Research Council (MRC)
Maximum ROM for each performance (degree)	MVIC testing
Angular velocity (degree/second) during going phase	FSHD-RODS
Angular velocity (degree/second) during returning phase	RWS
Average jerk (AJ, m/S3)	
Mean value of the RMS going phase (μV)	
Mean value of the RMS returning phase (μV)	
Muscle contribution (%) for each ROM	

RESULTS: Our device was utilized in a pilot study of healthy subjects and FSHD patients. The results of the maximum ROM during shoulder abduction, elbow flexion, and shoulder external rotation in both sides in four subjects are summarized in table 2. Maximum ROM for each subject is calculated as the average of 5 tries. In these results, the FSHD patient has overall lower ROM than the healthy subject for all three movements. For example, in flexion movement, left deltoid muscle contribution calculation in the FSHD patient subject 2was 21.96%, while in the healthy subject2 it was 38.2%. The angle value of maximum abduction in the left shoulder of the FSHD patient was 70.9 degrees and in the left shoulder of healthy subject was 119 degrees.

DISCUSSION: The objective of this study is to develop and test our new outcome measure that could be used to describe the central tendency and variablity of this test in FSHD patients as compared to healthy control subjects. After enrollment of FSHD or healthy subjects following informed consent, these baseline variables was first collected: participants demographics and anthropometric data (gender, age, weight, height, hand dominance), FSHD disease characteristics for FSHD patients (age of symptoms onset, disease duration, FSHD type), medical comorbidities, baseline status of assistive device use for ambulation, and status of physical therapy. The wearable sensors will then be used to measure shoulder girdle muscle activity and ROM outcomes. Our powered adjustable brace aims to reduce recovery time by giving real-time feedback to the user during postoperative rehabilitation, improve ROM and optimize pain control. Measurements from the device can be transmitted wirelessly to healthcare cloud/storage for further processing.

Figure 2. Maximum range of motion (ROM) measured by the wearable device.

	Subject	Gender	Age	Shoulder Abduction (deg)		Elbow Flexion (deg)		Shoulder External Rotation (deg)	
				Left	Right	Left	Right	Left	Right
Healthy	1	Male	29	108.6	120.6	165.1	147.2	67	54
	2	Female	74	119	179.4	133.5	165.3	72.2	78.9
FSHD	3	Male	31	81.7	91.4	132	123	19.1	43.7
	4	Female	63	70.9	48.8	92.9	157.2	63.2	30.3

SIGNIFICANCE:
In the healthcare cloud, an AI-based algorithm will estimate the impairment of individual shoulder girdle muscles. This technology can be used for remote at-home telemedicine services

REFERENCES: [1] Ghasemi M. et al. Cells 2022; [2] Geary, M.B. et al. Geriatr Orthop Surg Rehabil, 2015.