



## Case Report

# Acupuncture Improves the Facial Muscular Function in a Case of Facioscapulohumeral Muscular Dystrophy

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### Abstract

Facioscapulohumeral muscular dystrophy (FSHD) is a genetic muscle disorder in which muscles of the face, shoulder blades, and upper arms develop gradual and progressive weakness. There is no effective pharmacological treatment currently available for this disorder so far. We had an opportunity to treat a patient with FSHD using acupuncture. The patient was a 62-year-old female, who presented to us with symptoms such as weakness in her eyes, mouth, shoulder, and upper and lower limbs. Muscle atrophy could be found in multiple areas in her body including her face, shoulder, arm, chest, and lower limbs. Her diagnosis of FSHD muscular dystrophy was established a few years ago and was later genetically confirmed. After a long treatment course of about 10 months with acupuncture, this patient showed a significant restoration of her facial muscle function. However, acupuncture did not improve the function of other muscle groups. The potential mechanism that acupuncture improved the facial function but not the other muscles needs to be further investigated.

## 1. Introduction

Facioscapulohumeral muscular dystrophy (FSHD) is an autosomal-dominant slowly progressive muscular disorder found in adults, with a prevalence of 1: 15,000–1: 20,000 [1,2]. It is characterized by muscle weakness and atrophy,

which typically begin in the face and then migrate to shoulders and upper arms. Muscle weakness can also spread to the hip girdle muscles and muscles in the lower extremities, causing foot drop and walking difficulties. FSHD is slowly progressive and causes significant lifetime morbidity. Around 20% of patients will eventually become

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wheelchair dependent. There are currently no effective pharmacological treatments for FSHD [3]. Those suffering from FSHD often seek treatment from complimentary and alternative medicine. We had an opportunity to treat a patient with this disorder in the past two years and would like to report the treatment process and outcome herein.

## 2. Case description

Ms M. was a 62-year-old female, who presented to our clinic in May 2013, seeking acupuncture treatment for weakness in her eyes, mouth, shoulder, and upper and lower limbs. She had been diagnosed with FSHD muscular dystrophy a few years earlier and had genetic confirmation of her diagnosis. She had been taking estradiol, Centrum, and vitamin E on and off for years. She reported that her first symptom of muscle weakness appeared when she was 19 years old, although final diagnosis was not established for 30 years, in 2003. Genetic testing demonstrated that she carried a "de novo" D4Z4 reduced allele with 1 repeat (20 kb).

Patient's family history was positive for FSHD in her mother, grandmother, and two out of four siblings. Her elder brother was unaffected, and one of her elder sisters had not shown signs of FSHD, although she had unexplained mental retardation. Her mother and grandmother with FSHD had died of ovarian and breast cancer, respectively.

On physical and neurologic examination, the patient's vital signs were within normal limits. She appeared alert and awake, in no acute distress, and was pleasant and cooperative. There was marked weakness in the muscles controlling eye and mouth closure, as well as minimal function of muscles of facial expression. She could not frown and wrinkle her forehead; forehead lines and nasolabial grooves had completely disappeared. She could not close her eyes, pucker up her mouth, and puff up her cheeks. Obvious atrophy could be seen in the following muscles: the shoulder girdle muscle, supraspinatus, infraspinatus and subscapularis, serratus anterior, rhomboids, teres muscles, pectoralis major and minor, biceps and triceps, and brachioradial muscle. Bilateral scapular winging could be observed with normal strength to the neck flexion and extension. Both arms could not lift up more than 90°. Elbow flexion was normal on the left but weak on the right, whereas elbow extension was normal on the right but weak on the left. There was mild weakness of wrist extension bilaterally. Her right index finger could not extend but appeared to have normal strength of thumb and finger flexion. There was apparent weakness in leg muscles, and bilateral foot drop was evident. Although she had difficulty with ambulation, she was able to walk with an unsteady gait with the assistance of a cane. There was mild impairment of the hip abductor. Hip flexion was limited, weaker on the left than on the right. Knee extension was mildly impaired bilaterally, whereas flexion was heavily impaired. There was little movement for dorsiflexion or plantarflexion of the ankle bilaterally.

### 2.1. Treatment

Acupoint selection: Two groups of acupoints were selected. The first group of acupoints was used for the

patient in her prone position, whereas the second group was used for the patient while she was in her supine position.

The first group acupoints were UB-10, GV-14, UB-13 to UB-15, UB-18, and UB-20 to UB-25 bilaterally.

All needles were retained in place for 30 minutes, with manipulation at 10 minutes for each treatment.

The 2<sup>nd</sup> group of acupuncture points was ST-9, LI-4, SJ-9, LI-11, ST-36, GB-37, SP-6, EX-HN5, ST-7, ST-4, CV-24, ST-2, and GB-14 bilaterally.

All needles were retained in place for 20 minutes, with manipulation at 10 minutes for each treatment.

Needling method: A needle was inserted into UB-10 perpendicularly to a depth of 1.5 cun. Usually, a strong *Deqi* sensation could be produced in the head when the needle was inserted in that depth. A needle was inserted into UB-15 perpendicularly to a depth of 1.0 cun. A needle was inserted into ST-9 along the inner side of the common carotid artery to about 1.0–1.5 cun after pushing aside the sternocleidomastoideus muscle. A needle was inserted into GB-14 horizontally penetrating EX-HN4. Other points were needled routinely.

Treatment courses: The treatment was conducted three times a week. The time course for the treatment of this patient consisted of two episodes; the first one was 5 months in 2013, and the 2<sup>nd</sup> episode was another 5 months in 2014 (Owing to the cold weather in Minnesota, the patient could come to the clinic only during May to November). She was treated with 10 months of acupuncture all together.

## 3. Results

During the first three months of treatment (36 sessions), there was no subjective or objective improvement in muscle weakness in any part of the body, although the patient stated that she felt well with the treatments and wanted to continue. From the fourth month onward, the patient reported a mild improvement in eye closure during sleep. After 5 months of treatment, the patient and practitioner noticed a mild improvement in her facial expression. At this point, she appeared to have complete closure of her eyes.

The treatment was discontinued in November 2013 because of the cold weather and patient's inability to get to the center and resumed again in May 2014. When the patient came back for her treatment in May, we were surprised to observe that the improvement in eye closure and facial expression had sustained for 7 months. During the subsequent treatment cycle, strength in muscles of facial expression continued to improve as did the strength in her arms. Shoulder and lower extremities strength remained unchanged. At the end of this cycle of treatment, the patient was able to move her facial muscles much better. She could frown her eyebrow, close her eyes, and wrinkle her forehead without any difficulty. Her nasolabial groove became obvious. The orbicularis oris muscle function was much improved, and the patient could complete her pulmonary function test, which she was unable to perform during her physical examination in the last 10 years. Her arm force also had improved, although the weakness in the

**Table 1** The improved and unimproved muscular functions after acupuncture listed by the patient.

	Before	After
Face	No forehead wrinkles	Forehead wrinkles appear
	Eyes unable to close	Both eyes can close
	Eye droopy in the left side	Eye droopy disappears
	No top lip movement	Top lip movement is fine
	Flat droop on the left side when smiling	Flat droop is gone when smiling
Shoulder	Unable to suck on straw	Yes, can suck on straw
	Scapular wing	No change
Arms	Cannot lift more than 90 degree. Left arm (triceps) extension is weak; right arm (biceps) flex is weak.	No change
	Right pointer does not extend and cannot lift finger up	No change
Hand	Right pointer does not extend and cannot lift finger up	No change
Leg	Legs cannot flex, and hamstrings are weak.	No change
Foot	Adductions are very weak	No change
	Dorsiflexion is very weak	No change

shoulders and lower extremities remain unchanged even after the end of the 2<sup>nd</sup> cycle of treatments.

The patient herself listed a change in her facial, shoulder, arms, and legs before and after the acupuncture treatment, which is given in [Table 1](#).

#### 4. Discussion

FSHD was first described by Louis Landouzy and Joseph Dejerine in the late 1800s. Although major advances have occurred in the understanding of the genetics of this disorder over the past decades, the exact mechanism that leads to atrophy and weakness secondary to the genetic defect is still not well understood. Currently, there is no effective treatment for this disorder.

There is no similar terminology in Traditional Chinese Medicine (TCM), although description of muscle atrophy could be found in the earliest TCM book, *"The Yellow Emperor's Classics of Internal Medicine"* (黄帝内经). In the book, muscle atrophy was categorized as "Wei syndrome," and there was a special chapter in the book which illustrated the etiology (in TCM language) and treatment principle of muscle atrophy with acupuncture. According to this book, acupoints in the Yangming Meridians are always the first choice for acupuncture treatment in atrophy syndrome.

Besides, acupoints can also be selected according to a so-called Qi-Jie (Qi's pathway) theory. Its main concept is that the human body can be divided into four domains horizontally, the head, chest, abdomen, and low extremities. According to Lingshu, "The Qi in the head stores in the brain; the Qi in the chest stores in the chest muscles and the back-Shu points; the Qi in the abdomen stores in the back-shu points and the points in the Chong channels that lie around the umbilicus; and the Qi in the lower extremities store in the acupoint Qi-Chong (ST-30), Chengshan (BL-57), and the acupoints near the ankle" [4]. The acupoints lie in the area where the Qi stores are usually the most effective points for the treatment. Therefore, in this case, we select a couple of points from the back-Shu acupoints besides acupoints from the foot and hand Yangming meridians.

As there is no effective therapy for FSHD, this case report opens up a possibility that acupuncture may serve as an alternative modality for the treatment of this disease. However, both the practitioner and patient must keep in mind that the treatment may need a relatively longer period of time before seeing any significant improvements as most FSHD cases are of chronic onset and have a chronic development. In this case, a mild improvement in the facial muscle was only seen at the fourth month of acupuncture treatment (after 50 sessions of acupuncture). It is also interesting that effects of acupuncture can be long lasting, as seen with ongoing treatment response in our patient despite a 7-month hiatus in treatment.

The mechanistic study of acupuncture treatment of muscular atrophy is still lacking. Some studies showed that acupuncture stimulated the release of insulin-like growth factor-1 [5,6,7], a beneficial factor for the muscle regeneration. Also, the adenosine released around the needling area of the muscle could be converted into ATP [8,9,10], which could participate in the cellular respiration in the muscle and promote the cell regeneration or rescue the cells. However, it is still difficult to understand why acupuncture worked only in the muscles of the face and not other parts of the body in our patient. Therefore, an experimental study is needed to explore the potential mechanism of acupuncture in restoring the muscle function.

#### 5. Conclusion

We describe application of acupuncture to successful treatment of a patient with FSHD. After treatment of 10 months in 2 cycles, our patient's facial muscular function was almost completely restored, and her arm muscle weakness had also improved. There was no improvement in the shoulder and scapular muscles or in muscles in the hip and lower extremities. Based on the results of our patient, we propose that acupuncture could be considered as an ancillary treatment modality to help improve muscle function in patients with FSHD, especially those with facial muscle dystrophy. A future clinical trial is warranted to study more precise benefits, side effects, and application

of acupuncture in FSHD and to define clinical criteria for acupuncture use.

## Contribution

Y.L. performed the treatment. Y.L., F.X., and X.L. discussed the treatment protocol and evaluated the result. X.L. wrote the article.

## Disclosure statement

The authors declare that there are no conflicts of interest and no financial interests related to the material of this manuscript.

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