Kaohsiung J Med Sci 16: 248 — 254, 2000

THE STRENGTH AND FUNCTIONAL PERFORMANCE IN PATIENTS WITH FACIOSCAPULOHUMERAL MUSCULAR DYSTROPHY

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Facioscapulohumeral muscular dystrophy (FSHD) is a slowly progressive myopathy with autosomal dominant inheritance remarkable for its early involvement of facial musculature. The purpose of our study was to assess the rate of strength deterioration, functional condition and performance of activity of daily living of patients with FSHD in Taiwan. Twenty patients diagnosed with FSHD were included in this study. Manual muscle testing (MMT) was used to evaluate muscle strength. The Brooke and Vignos scales were used to assess upper and lower extremity function respectively, and the capability of the activity of daily living was measured by Barthel index. The result of the strength testing was characterized by the presence of a progressive asymmetrical muscular weakness in patients with FSHD. The mean muscular strength of the right extremity was weaker than its left counterparts ($p<0.05$) and the shoulder muscle group was the weakest. According to the Brooke functional scale, 20% of our patients were graded as 1, 30% as grade 2, and 50% as grade 3. On the Vignos functional scale, 50% of patients fell into grade 1, 10% in grade 2, and 40% in grades 3-5. Vignos scale was significantly correlated with mean muscle strength ($p<0.05$). The average value of Barthel index was 97.8 ± 4.7. The muscle strength decline in this Taiwanese of FSHD population was more severe in shoulder girdle area. The mean muscle strength of the right extremity was weaker than the left. Most of our patients suffered from mild or moderate physical disability. Finding of these Taiwanese FSHD population is similar to those reported elsewhere in the world.

Key words: facioscapulohumeral muscular dystrophy, manual muscle testing, functional grading

(*Kaohsiung J Med Sci* **16**: 248 — 254, 2000)

Facioscapulohumeral muscular dystrophy (FSHD) is a slowly progressive myopathy with an autosomal-dominant inheritance trait. The disease process is slow progressive myopathy. Facial weakness is often the earliest detectable sign, followed by muscle weakness in one or both shoulder girdles. Gradually, the foot extensors and muscles of the trunk and pelvic girdle become involved [1]. Most of the patients have greater involvement of proximal musculature, although a subgroup demonstrates early weakness of the ankle dorsiflexors [2]. This disease was noted for the very wide range of age at first symptomatic onset. This could vary from infancy to late adulthood, and showed inverse correlation with eventual severity, onset age was youngest and severity was greatest in isolated cases [3].

As FSHD is a disease of progressive muscular weakness, assessing the rate of deterioration of

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Received: January 10, 2000 Accepted: April 20, 2000

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strength was a critical tool used in therapeutic trials or evaluation of the natural outcome. The natural history and variable patterns of clinical progression for FSHD have not been well delineated. Muscle strength and disability had been simply studied in patients with FSHD in many countries, such as United Status, Japan, England, Netherlands and Russia in 1995-1996 [2, 4-6]. There is still no detailed strength and function evaluation of patient with FSHD in Taiwan population. Manual muscle testing was an established, reliable method to evaluate strength in neuromuscular disease and the commonly used Brooke and Vignos functional scale were simple tests for rating the motor function of upper and lower extremities [7,8]. Assessment of independence in basic functions of daily living for the disabled person has been accomplished in rehabilitation medicine using standardized scales, such as the Barthel index [9,10], which we also used to evaluate the activity of daily living (ADL) in FSHD patients. The purpose of this study was to assess muscle strength, extremity motor functions and status of independence in activities of daily living in the FSHD population in Taiwan.

MATERIALS AND METHODS

Twenty FSHD patients (12 males and 8 females) with a mean age of 35.5±16.6 years (ranging from 13 to 80 years) participated in this study. Their average age of onset was 18.2 ± 7.4 years, ranging from 9 to 30 years. The average disease duration was 17.3 ± 14.5 years, ranging from 4 to 70 years. All FSHD patients were diagnosed and followed at least for three years within a national multi-center registration program in Taiwan. Four main criteria were used to define FSHD: (1) onset of the disease in facial or shoulder girdle muscles, sparing of the extraocular, pharyngeal and lingual muscles and the myocardium; (2) facial weakness in more than 50% of the affected family members; (3) autosomal dominant inheritance in familial cases; and (4) evidence of myopathic disease in electromyography and muscle biopsy in at least one affected member, without biopsy features that are specific to alternative diagnoses. Disorders such as Duchenne and Becker muscular dystrophy, spinal muscular atrophy, mitochondria and metabolic myopathies were excluded. These diagnostic criteria were adopted from the new criteria of the European neuromuscular Community [11].

After medical and neurologic examination and confirmation of diagnosis by two neurologists, the patients were evaluated by a registered physical therapist. The physical therapy evaluation included the following: (1) Manual Muscle Testing (MMT) for strength measurement, (2) the Brooke and Vignos scale for assessing the function of the upper and lower extremities and (3) the Barthel index for measuring ADL performance.

In this study MMT included the strength measurement of neck/trunk and upper/lower extremities. In all, 32 (fourteen per side of upper/lower extremities, four groups of neck and trunk) muscle groups were examined. Those muscle groups were shoulder flexors/extensors/abductors, elbow flexors/extensors, wrist flexors/extensors, hip flexors / extensors / abductors, knee flexors / extensors, ankle dorsiflexors/plantaflexors, neck flexors / extensors, trunk flexors / extensors. The grading methods for muscle strength, using gravity as resistance to grade strength (grade as: Zero, Trace, Poor, Fair, Good and Normal). Note that a grade of Normal represents normal strength and a grade of Zero represents no contraction felt. For computational purposes, the grading system has been translated to as follows: Normal grade as (5), Good grade as (4), Fair grade as (3), Poor grade as (2), Trace grade as (1) and Zero grade as (0). The minus and plus symbols would translate as follows: Fair + as (3.33), Fair – as (2.66), Poor + as (2.33) and Poor – as (1.5) [12].

The Brooke Scale was applied for grading the upper extremities function [7] and the Vignos scale for the lower extremities [8]. Table 1 shows a description of Brooke and Vignos scales. The Barthel index was used to measure the ADL performance [10]. The total scores were broken down into the following severity categories: 0-20 was totally, 21-61 severely, 62-90 moderate and 91-99 mildly dependent. A score of 100 was regarded as totally independent. This system was simple and easy to use and has been proven to be an effective index of severity of ADL dependency [10].

The strength of 32 muscle groups was averaged and named as the "mean muscle strength". To investigate understand the difference of left and right side strength, and the upper and the lower extremities strength, average left and right side extremities strength and average upper and lower extremities strength were calculated. Paired t-test was used to compare the side differences between the mean extremities strength, flexor/extensor muscle strength of each joint and average upper/lower extremities strength. We also explored the relation between strength deterioration, motor functional condition, ability of ADL and age/disease duration. The Pearson correlation coefficient was used to correlate mean muscle

Table 1. Grading for Brooke and Vignos functional scales

Grade	Functional description
Brooke scales for upper extremity	
1	Starting with arms at the sides, the patient can abduct the arms in a full circle until they touch above the head
2	Can raise arms above head only by flexing the elbow (shortening the circumference of the movement) or using accessory muscles
3	Cannot raise hands above head, but can raise an 8-oz glass of water to the mouth
4	Can raise hands to the mouth, but cannot raise an 8-oz glass of water to the mouth
5	Cannot raise hands to the mouth, but can use hands to hold a pen or pick up pennies from the table
6	Cannot raise hands to the mouth and has no useful function of hands
Vignos scales for lower extremity	
1	Walks and climbs stairs without assistance
2	Walks and climbs stair with aid of railing
3	Walks and climbs stairs slowly with aid of railing (over 25seconds for eight standard steps)
4	Walks unassisted and rises from chair but cannot climb stairs
5	Walks unassisted but cannot rise from chair or climb stair
6	Walks only with assistance or walks independently with long leg braces
7	Walks in long leg braces but requires assistance for balance
8	Stands in long leg braces but unable to walk even with assistance
9	Is in a wheelchair
10	Is confined to a bed

strength and Barthel total score, mean muscle strength and age/disease duration, Barthel total score and age/disease duration. The Spearman correlation coefficient was used to determine the relationship between the Brooke and Vignos functional grade, Barthel total score, mean muscle strength and age/disease duration. Statistical significance was set at $p<0.05$.

RESULTS

The demographic data and the results of physical therapy evaluations of the FSHD patients are presented in Table 2. On average, the "mean muscle strength" of our patients was 3.77 ± 0.56. The shoulder muscle groups were the weakest, with mean strength between 2.77-2.78. Elbow muscle groups followed these, with mean strength ranging between 3.64-3.65. The neck muscle groups were 3.68-3.73. Other muscle group, such as trunk and lower extremities, mean strengths were between 3.95-4.15. The least impaired muscle groups were wrist muscle groups, with a mean strength of 4.22-4.35. Average upper extremities muscle strength was significantly weaker than average lower extremities muscle strength ($t = 3.98$, $p<0.001$). Average right side muscle strength was significantly weaker than average left side muscle strength ($t = 2.85$, $p<0.05$). Mean muscle strength was not significantly correlated to age or disease duration ($r = -0.284$, -0.177; $p>0.05$).

The function of upper and lower extremities was graded on the Brooke and Vignos scales respectively. Four patients (20%) fell into grade 1 category. Six (30%) were graded as 2; and 10 (50%) were graded as 3. There was no patient in grades 4-6. The function of lower extremities was graded on the Vignos scale: ten (50%) in grade 1, 2 (10%) belonged to grade 2, 3 (15%) to grade 3 and 4 (20%) patients in grade 4. There was only one (5%) patient in grade 5. No patient was in grades 6-10. Table 3 represents the results of Brooke and Vignos scales. The Brooke scale was not significantly correlated to age or disease duration ($r = 0.254$, 0.355; $p>0.05$). The Vignos scale was also not significantly correlated to age ($r = 0.301$, $p>0.05$) but was significantly correlated to disease duration ($r = 0.483$; $p<0.05$). The relationship of motor function with mean muscle strength showed that the Brooke scale was not significantly correlated to mean muscle strength or average upper extremity muscle strength ($r = -0.052$, -0.339; $p>0.05$). The Vignos scale was significantly correlated to mean muscle strength and average lower extremity muscle strength ($r = -0.583$, -0.536; $p<0.05$).

Table 2. Simple descriptive statistics of patients with facioscapulohumeral muscular dystrophy

	Total patients	Male	Female
Number	20	12	8
Age	35.5 ± 16.6 (13-80)	35.8 ± 19.2 (19-80)	35.0 ± 13.2 (13-54)
Age of onset	18.2 ± 7.4 (9-30)	17.8 ± 7.4 (10-30)	18.6 ± 8.0 (9-30)
Disease duration	17.3 ± 14.5 (4-70)	17.9 ± 18.0 (4-70)	16.4 ± 7.7 (4-27)
Mean muscle strength	3.77 ± 0.56 (2.72-4.72)	3.85 ± 0.67 (2.72-4.72)	3.65 ± 0.37 (3.01-4.22)
Barthel index	97.8 ± 4.7 (85-100)	99.2 ± 2.9 (90-100)	95.6 ± 6.2 (85-100)

Mean ± SD (range)

Table 3. Distribution of grades for motor function

Type of scale	Number (%)
Brooke scale	
1	4 (20)
2	6 (30)
3	10 (50)
Vignos scale	
1	10 (50)
2	2 (10)
3	4 (20)
4	3 (15)
5	1 (5)

Mean total score of the Barthel index for ADL was 97.8 ± 4.7 (Table 4). Sixteen patients (80%) received full score (100), three (15%) scored 90 and one scored 85. According to ADL performance assessed in degrees of severity, 16 (80%) patients were classified as totally independent, 4 (20%) patients were moderate dependent. Of all ADL activities, the up and down stairs needed most assistance (average score=8.5, full score=10), this was followed by bathing (average score=4.75, full score=5) and dressing (average score=9.5, full score=10). Other items, such as feeding activity, personal toilet, bowel/bladder control ability, transfer ability, and ambulation were totally independent. The Barthel total scale was not correlated to age or disease duration ($r = -0.398, -0.212$; $p>0.05$), but was significantly correlated to mean muscle strength ($r = 0.474$, $p<0.05$).

DISCUSSION

In the present study, the pattern of declining strength, motor function status and ADL was analyzed in 20 FSHD patients. The results showed that the mean muscle strength of these patients sustained mild to moderate decreases. Shoulder muscle groups were weaker than any other muscle groups, with mean strength between 2.77-2.78 (translated from MMT scores, not actual strength by machine). In the "fair -" grade muscle strength equal to 2.66, with such shoulder muscle strength, the arm could only be raised slightly against gravity but could not complete the arc of motion. The order of severity of strength decreased was shoulder then elbow then lower extremity muscle then trunk and finally wrist muscles. In a "the FSHDY group" population the weakest muscle group was also reported to be the shoulder muscle groups [13]. Kilmer *et al.* studied 22 subjects with FSHD revealed that extensor muscles were significantly weaker than flexors, proximal muscles weaker than distal, and ankle dorsiflexors weaker than the plantar flexors [2]. In our patient population we did not find that extensor were weaker than flexors ($t = -1.422$ to 1.283; $p>0.05$), but

Table 4. The dependent condition of Barthel score

Severity categories of dependence	Barthel score	Number	Percentage
Total independent	100	16	80%
Moderate dependent	90	3	15%
	85	1	5%

ankle dorsiflexors (3.95) was weaker than the plantarflexors (4.15) ($t = 2.629$, $p<0.05$).

Asymmetry of upper extremity musculature has been reported previously. Some of these studies showed greater weakness of selected dominant limb muscle groups due to overuse [5, 14]; others disagreed [2, 15]. The right side mean muscle strength in our subjects was weaker than the left side ($t = -2.854$, $p<0.05$). This finding is supportive of the relation between handedness and increased muscle weakness in right-handed FSHD patients, suggesting that mechanical factors such as overuse might play a distinct role in the progression of muscle weakness in FSHD [5]. Tawil noted that major right/left side-to-side differences in strength but these were not related to handedness [15]. In Chinese culture, parents trained their children to use right hand for eating, writing and most of their daily living activities. We suggested that muscular weakness of the right extremities in FSHD might get worse by overuse or overload exercise in Taiwanese FSHD population. Upper extremity muscles (shoulder external rotators, shoulder abductors, and wrist extensors) were also reported to be significantly weaker in dominant limb [2]. Asymmetry in muscle strength in our subjects is such that the right side was weaker than left side and occurred both in the upper and lower extremities, more often on the muscles of the shoulder, hip and ankle.

Some studies reported the clinical heterogeneity in the FSHD population [2, 4, 6]. The muscle weakness pattern of subjects with FSHD had a proximal predilection as measured by MMT. Exceptions were those with significant weakness of the ankle dorsiflexors [2]. They found particular involvement of the peroneal musculature in certain patients, and suggested a "facioscapuloperoneal" variation [6,16]. Although the ankle dorsiflexor muscle strength of our patients was slightly weaker than plantaflexor, ankle dorsiflexor was not as severely affected as shoulder muscles. A subgroup of FSHD population (18%) reported by Kilmer *et al.* showed that the ankle dorsiflexors were at least one grade weaker than the hip extension [2]. We were not able to identify any facioscapuloperoneal type of muscular dystrophy in our FSHD population. Scapuloperoneal syndromes with variable degrees of facial weakness could occur in a substantial number of patients with symptoms that mimic those of FSHD. Different gene locations between FSHD and scapuloperoneal syndrome have been reported [17]. Some studies suggested genetic heterogeneity for the FSHD patients such as in British and Dutch populations [18, 19], but others studies did not suggested any genetic heterogeneity for the patients in the Brazilian, Japanese and Taiwanese populations [20-22].

The mean muscle strength did not significantly correlate to age or disease duration in our FSHD subjects. Many studies found that an early age of onset was associated with greater likelihood of more severe weakness [2-4, 13]. Kilmer et al. reported that an age of onset earlier than 15 years had an average MMT score of 3.0 units or less [2]. The wide range of severity in FSHD complicated the clinical practice of genetic advice, although onset age is youngest and severity is greatest in isolated cases. They propose that quantitative variation in a uniform mutation mechanism influencing age at onset, but by deletion rather than expansion of DNA [3]. Fragments in the smallest size range (13-18 kb) result in the earliest onset and greatest clinical severity. Larger cosegregating fragments (>30 kb) could be associated with a later onset and milder presentation of FSHD. Patients in our study, the age of onset was late (mean $\pm$ SD: 18.15 ± 7.44, range 9-30). The onset age of most of our subjects was 10-20 years old (50%) and 21-30 years old (45%). We could not confirm the gene deletion condition in this study, but there was no correlation between onset of age and severity of this disease in our patients.

The Brooke scale and the Vignos scale were used to evaluate upper and lower extremities motor function respectively. All of our patients, Brooke scale was 1-3 grade (20% in grade 1, 30% in grade 2, 50% in grade 3). Similar results were reported in a FSH-DY group [13]. Kilmer *et al.* found 32% of patients in Brooke grade 1 could abduct their arms in a full circle above their head and 95% of patient's upper extremities functional grade was between grade 1 and 3 [2]. Upper extremity function in FSHD patients showed only mild impairment, primary on shoulder muscle. They all had the ability to raise a glass of water to the mouth by elbow flexion movement. The lower extremity motor function of Vignos scale showed that 50% of patients, who were in grade 1, could walk and climb stairs without assistance. Kilmer *et al.* found that in such population, 48% could walk and climb stairs without assistance (grade 1), 38% in grades 2-3, and 14% in grade 9 were unable to walk and used a wheelchair [2]. The FSH-DY group study also found about 9% of the patients need to use a wheelchair [2]. No patient was unable to walk and needed wheelchair in our subjects. More patients with FSHD or continue follow up studies after 5-10 years may change the percentage of patients in wheelchair condition. Patients with FSHD after confined to wheelchair were more difficult to visit hospital. This difficulty to access hospital

may be the reason that better lower extremity function were found in our patients population in Taiwan.

The Barthel index showed a high score for capacity of ADL in patient with FSHD. Eighty percent of patients had the full score (100). Comparing to Duchenne or limb-girdle type muscular dystrophy, the FSHD subjects showed a better ability of daily living activity measured by Barthel index [23, 24]. In spite of upper limb weakness, and reported difficulties with ADL, they used the contralateral arm to elevate the elbow so that the hand could reach the top of the head to comb their hair or perform other activities [25]. The item of dependent activities was up/down stairs, bathing and dressing. The Barthel index might not be appropriate for evaluating ADL for patient with FSHD. A more detailed measurement system was needed to evaluate the ability of daily living performance for the mild form of muscular dystrophy.

In summary, the FSHD population investigated in the present study showed greater involvement of the shoulder musculature. The mean muscle strength of the upper extremity was weaker than that of the lower extremity and the right extremities were weaker than their left counterparts. More studies are needed to investigate whether strengthening exercises and overuse are detrimental to FSHD patients. The scores of extremity motor function testing with Brooke and Vignos scaled ranged from 1-3 for the upper extremities and 1-5 for the lower extremities. The activities of daily living showed only mild impairment. Eighty percent of patients in our FSHD population are totally independent in ADL. More challenging tasks were climbing up/down stairs, bathing and dressing. Environmental modification and the use of assistant devices may be beneficial for these patients.

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顏肩肱型肌肉失養症病人之肌力與功能表現

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顏肩肱型肌肉失養症（facioscapulohumeral muscular dystrophy）是屬於遺傳性緩慢漸進性肌肉萎縮無力之疾病，遺傳方式以常染色體顯性遺傳並發生臉部肌肉之無力。本篇的研究目的為研究分析顏肩肱型肌肉失養症病患自然疾病過程之中，肌力喪失情形與運動功能狀態以及日常生活能力之表現等。病患共有 20 人，以徒手肌力測試評估上下肢肌力狀態，以 Brooke 和 Vignos 評估表來評估上下肢運動功能，以巴歇爾指數（Barthel index）來評估日常生活能力。結果發現顏肩肱型肌肉失養症病人肌力的下降呈現漸進式且不對性肌力衰退，肩關節的肌力是肢體肌力最差的關節，病人常出現左右兩側肌力不等的情形，右側肌力低於左側肌力($p<0.05$)。上肢運動功能分佈之等級分別為等級1佔20%，等級2佔30%以及等級3佔50%。下肢運動功能分佈之等級分別為等級1佔50%，等級2佔10%以及等級3-5佔40%。下肢運動功能與下肢肌力有顯著相關($p<0.05$)。平均巴歇爾指數分數為 97.8 ± 4.7 。本篇研究提供台灣顏肩肱型肌肉失養症病患，自然疾病過程肌力之下降情形與功能表現，可作為臨床治療肢帶型肌肉失養症病患的參考。

(高雄醫誌 **16**: 248 — 254, 2000)

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收文日期：89 年 1 月 10 日　　接受刊載：89 年 4 月 20 日
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