ORIGINAL ARTICLE

# Activities of Daily Living (ADL) Structure of Patients with Duchenne Muscular Dystrophy, Including Adults

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**Abstract**

***Objective*: To study the activities of daily living (ADL) structure of patients with Duchenne muscular dystrophy (DMD).**

***Design*: Cross-sectional survey.**

***Subjects*: Seventy-two patients with DMD (mean [SD] age, 17.2 [8.1] years), recruited in 160-bed regional center for neuromuscular disease.**

***Methods*: We assessed 72 participants with the Functional Independence Measure (FIM$^{SM}$), and determined the difficulty order of the FIM$^{SM}$ items with the percentage of patients who were independent (6 or 7) for each FIM item (% independence).**

***Results*: The ADL of patients with DMD had an order in terms of difficulty. For the motor subscale, the most difficult item was stairs, and the easiest item was locomotion. On the cognitive subscale, problem solving was the most difficult item. When we compared item difficulty between patients <15 years of age and those ≥ 15 years, the % independence was lower in the older age group for all motor items. The decrease in the % independence of the eating item with age was significant; however bladder management was well maintained.**

***Conclusions*: ADL of patients with DMD has a specific difficulty order. The order of difficulty is based on the relative level of independence/dependence. The difficulty order was maintained, although the structure of ADL changed with age.** (Keio J Med 58 (4) : 223–226, December 2009)

***Keywords*: activities of daily living, muscular dystrophy, functional independence measure**

## Introduction

Duchenne muscular dystrophy (DMD) is an X-linked recessive dystrophic myopathy caused by complete absence of the protein dystrophin in the skeletal muscle, myocardium, and brain. Patients develop progressive weakness, contractures, spinal deformity, restrictive lung disease, cardiomyopathy, and variable degrees of cognitive impairment - all contributing to progressive impairment in the ability to complete activities of daily living (ADL). With advances in respiratory management, life expectancy of patients with DMD has increased, and the mean age at death has increased from 14.4 years in the 1960s to 25.3 years in the 1990s.[1] Despite this improvement in survival, disease progression is inevitable, leading to a state of severe physical dependence. Therefore, one of the important goals of patient management is the maintenance of ADL as long as possible. However, only a few reports investigating ADL in patients with DMD have used valid and reliable instruments,[2,3] and no study includes patients aged 15 and older, despite the fact that many patients now survive into adulthood. The aim of this study was to use the Functional Independence Measure (FIM$^{SM}$),[4,5] a validated functional instrument, to assess disabilities in patients with DMD over a wide age range and to analyze the percent of independence associated with each FIM$^{SM}$ item.[3]

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## Methods

Participants were recruited from male inpatients and outpatients of a 160-bed regional center of neuromuscular diseases in the Saitama Prefecture in the northern part of the Tokyo Metropolitan area with a population of approximately 7 million. The number of patients with DMD in this area is estimated to be 240 based on the incidence of DMD (6.3/100,000 live births) and the number of males (3.5 million) in the prefecture. The diagnosis of DMD was established with clinical criteria, family history, dystrophin analysis of muscle biopsy specimens, and gene analysis by child neurology specialists. We selected patients aged 7 and older. We excluded patients with acute illness or other conditions that would prevent us from obtaining informed consent. Female carrier cases were excluded.

The institutional ethical committee approved the study, and informed consent was obtained from patients or family members before data collection in accordance with the Declaration of Helsinki.

Three physiatrists trained and experienced in the instrument assessed the FIM$^{SM}$ both by interviewing the parents or nurses and by directly observing the patients' performance. We also calculated the percentage of patients who were independent (scored at 6 or 7) for each FIM item (% independence), which was used as an indicator of item difficulty in a previous study.[3] Furthermore, we compared item difficulty, indicated as % independence, between patients aged ≥15 years and those <15 years.

## Results

Seventy-two male patients with DMD were recruited for this study. There were 35 inpatients and 37 outpatients (mean [SD] age, 17.2 [8.1] years, range 7 to 40 years, median age, 15.5 years). Twenty-nine patients used electrical powered wheelchairs, 19 patients used manual wheelchairs, 12 patients used assisted type wheelchairs, and 12 patients were ambulatory. Fifteen patients used a ventilator part-time and 7 patients used a ventilator 24 hours a day; ventilators were not needed for the remaining 50 patients. The FIM$^{SM}$ motor scores ranged from 14 to 91, and the mean (SD) and median scores were 37.3 (18.1) and 33, respectively. The FIM$^{SM}$ cognitive scores ranged from 11 to 35, and the mean (SD) and median scores were 32.0 (5.0) and 35. The FIM$^{SM}$ motor score significantly correlated with age (Spearman's rank correlation method [Rs= -0.66, $p$<0.001]) (**Fig. 1**). The mean FIM$^{SM}$ motor score for patients aged <15 years (n=34) and those ≥15 years (n=38) were 47.4 and 28.3, respectively ($p$<0.001, Mann-Whitney U test).

The FIM$^{SM}$ motor score was analyzed by the type of locomotion: ambulatory (n=12), manual wheelchair (n=19), electrical powered wheelchair (n=29), and assisted type wheelchair (n=12). The mean FIM$^{SM}$ motor score for each type of locomotion was 66.6, 38.7, 31.2, and 20.5, respectively ($p$<0.001, Kraskal-Wallis test). In patients ≥15 years, the mean FIM$^{SM}$ motor score of ventilator users (n=22) was significantly lower than that of non-ventilator users (n=16) (25.2 vs. 32.5; $p$<0.001, Mann-Whitney U test).


**Fig. 1** The relationship between FIM motor score and age.
The correlation between the FIM motor score and age was statistically significant with Spearman's rank correlation method (Rs= -0.66, $p$<0.001).

**Table 1** shows the % independence of FIM items. The most difficult item was stairs, and only 5 patients scored at independent levels of 6 and 7. Easier items included locomotion (the easiest item), bowel management, and eating. Regarding the locomotion item, 60 of 72 patients (83.3%) used wheelchairs and 41 of those 60 (68.3%) scored 6. As a rule, ADL items related to lower extremity functions (e.g., transfer items and dressing the lower part of the body) were relatively difficult, whereas items related to upper extremities (e.g., eating, grooming, bathing, and dressing the upper part of the body) were easier.

When comparing the item difficulty between patients <15 years and those ≥15 years, the % of independence was lower in the older age group for all motor items. The % independence of eating items was 7.8 in ≥15 group but 76.4 in <15 group. The decrease in % independence was biggest in the eating item and smallest in the bladder management item. Chi-square test showed no significant difference of % independence between the ≥15 years and <15 years groups in bladder management, bowel management, comprehension, and expression items.

**Figure 2** illustrates the percentage of patients requiring assistance (FIM$^{SM}$ score ≦4), supervision (FIM$^{SM}$ score=5), and those who were independent (FIM$^{SM}$ score ≧6) for each FIM$^{SM}$ item.

With regard to cognitive items, the % independence of problem solving was the lowest (i.e., it was the most difficult item). The % independence for problem solving item was low in patients <15 years, but not in those ≥15 years. The % independence of memory, social interac-

**Table 1** Item difficulties of the FIM

| Items | % of independence (total) | % of independence(<15) | % of independence( ≧ 15) | Chi-square |
|---|---|---|---|---|
| **Motor** | | | | |
| Locomotion | 66.6 | 88.2 | 65.7 | $p$=0.025 |
| Bowel | 63.8 | 76.4 | 55.2 | $p$=0.059 |
| Eating | 40.2 | 76.4 | 7.8 | $p$<0.001 |
| Bladder | 23.6 | 26.4 | 21.0 | $p$=0.589 |
| Grooming | 15.2 | 26.4 | 5.2 | $p$=0.003 |
| Bathing | 12.5 | 23.5 | 2.6 | $p$=0.007 |
| Dressing U | 15.2 | 32.3 | 0 | $p$<0.001 |
| Toilet | 9.7 | 20.5 | 0 | $p$=0.003 |
| Toilet transfer | 12.5 | 26.4 | 0 | $p$=0.001 |
| Dressing L | 11.1 | 23.5 | 0 | $p$=0.002 |
| Bath transfer | 11.1 | 23.5 | 0 | $p$=0.002 |
| Bed transfer | 9.7 | 20.5 | 0 | $p$=0.003 |
| Stairs | 6.9 | 14.7 | 0 | $p$=0.004 |
| **Cognition** | | | | |
| Expression | 87.5 | 91.1 | 84.2 | $p$=0.372 |
| Memory | 88.8 | 79.4 | 97.3 | $p$=0.016 |
| Social | 90.2 | 82.3 | 97.3 | $p$=0.03 |
| Comprehension | 73.6 | 73.5 | 73.6 | $p$=0.98 |
| Problem | 72.2 | 52.9 | 89.4 | $p$=0.001 |

Bowel; bowel managementl, Bladder; bladder management, Dressing U; dressing upper body, Toilet; toileting, Dressing L; dressing lower body, Social; social interaction, Problem; problem solving.
The difference of % of independence between <15 and ≧15 was statistically analyzed with chi-square test.

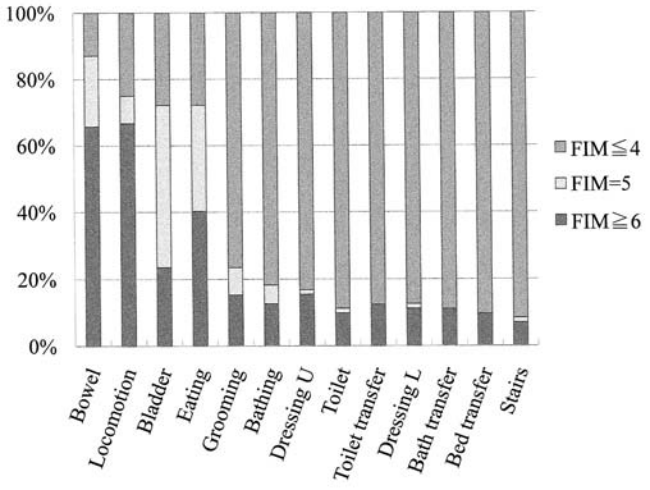


**Fig. 2** Percentages of patients requiring assistance (Functional Independence Measure (FIM$^{SM}$) score ≦4), supervision (FIM$^{SM}$ score = 5), and who were independent (FIM$^{SM}$ score ≧6) for each FIM$^{SM}$ item.

tion, and problem solving items were significantly higher in those ≥15 years than in those <15 years (chi square test, $p$=0.016, 0.03, and 0.001, respectively).

## Discussion

Few studies use validated instruments to examine the ADL of persons with DMD. Nair, *et al.*[2] used the Barthel Index[7] to assess ADL in 31 children with DMD, and our group[3] measured the FIM$^{SM}$ in 27 children. The FIM$^{SM}$ is a standardized functional instrument widely used to assess various disabling conditions. This study assessed FIM$^{SM}$ of patients with DMD including those < 15 years and ≥15 years, the latter of which have not been included in previous reports. The FIM motor score was negatively correlated with age. The difference in the FIM motor score between patients <15 years and those ≥15 years was statistically significant. The relative stability of the FIM motor score in patients ≥15 years indicates that the level of assistance required is relatively stable from the later stages of the second decade on. It appears that some FIM domains, such as locomotion, are relatively insensitive measures of disease severity because of the use of power wheelchairs to maintain modified independence.

In terms of item difficulties, Nair, *et al.*[2] analyzed the prevalence of partial or total dependence among Barthel Index items, and our group[3] assessed them by calculating the percentage of patients requiring assistance. In our previous study,[3] eating was the easiest item, followed by bowel management and locomotion. In the current study, locomotion was the easiest item, followed by bowel management and eating. Eating item ranked third from

the easiest. This discrepancy could be explained by the difference in age distribution, which ranged from 7 to 40 years in the present sample and from 7 to 14 years in our previous sample. The progressive decline in muscle strength that occurs with aging is likely to have rendered more patients to become dependent in eating, as evidenced by the smaller % independence for eating in the present study compared with our previous study (38.8% vs. 85.1%). This finding was confirmed by performing % independence analysis separately for the two age groups.

For the bladder management item, the % independence was 23.6%, whereas the median item score was 5. This finding could be due to the fact that many patients maintained the voiding sensation and intact bladder function, while at the same time they needed assistance to set or hold their urinal.

Locomotion was listed as one of the easier items in this study. This is because many patients maintained their capability for locomotion –even with advancing age– with the help of an individually tailored electrical wheelchair equipped with a seating system and a non-invasive ventilator.

Among the three transfer items, toilet transfer was the easiest and bed transfer was the most difficult. This could be related to proximal weakness of the lower extremities, because it is necessary for patients to lift their feet while performing bed transfers but not while performing toilet transfers.

DMD is a progressive disease in which the independence of ADL decreases with time. Our study showed the decrease of % independence for each FIM item was not always correlated with its % independence value. For example, in patients <15 years, the % independence of grooming and toilet transfer items were the same. However, in subjects ≥15 years, the % independence of grooming was higher than that of toilet transfer. It is also possible that the cross-sectional nature of this study and the small sample size were not enough to establish a task difficulty hierarchy in younger and older age groups. Future studies with larger sample sizes and longitudinal data collection are warranted.

Our study also indicated that there was a difficulty order in the cognitive items of the FIM$^{SM}$. In this population, an intellectual impairment has been documented,[7] and an early non-progressive impairment of verbal intelligence and mental retardation are reported.[8] Our study showed that problem solving was the most difficult item, for which more than 20% of the patients required some assistance. The % independence of problem solving was lower in patients <15 years than those ≥15 years. It is likely that it is more difficult for younger children to accept their impairment and disability. It is therefore necessary to consider more mental care for these younger patients. Problems related to verbal impairment and attentional complexities are known to occur in DMD.[8] When mental retardation is present, motor development landmarks could be slowed, and the children are likely to cease ambulation earlier.[7] This, in turn, could influence educational placement. We therefore recommend paying attention not only to motor function but also to cognitive function when helping patients with DMD manage their ADL.

The major goals of rehabilitation for patients with DMD are maintenance of ADL and the development of a program for prevention of progression of disability as well as supportive counseling for patients and families.[7] It is therefore necessary to know the level of difficulty associated with each ADL and how ADL declines with age. The results of this study might be useful in predicting the progression of difficulties in completing ADL for patients with DMD and to help form a plan to maintain ADL as long as possible.

## Conclusions

This report demonstrated that ADL of patients with DMD had a specific order of difficulty. The order of difficulty in this study is slightly different from previous studies, whose subjects were younger.

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