

Effect of Activities of Daily Living on Quality of Life among Japanese Adults with Duchenne Muscular Dystrophy: A Cross-sectional Study

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Abstract

Objectives: This study aimed to evaluate the relationship between activities of daily livings (ADLs) and psychological distress along with the quality of life (QOL) in Japanese adults with Duchenne muscular dystrophy. **Patients and Methods:** This study surveyed 36 adults using the Functional Independence Measure (FIM), the 36-Item Short-Form Health Survey (SF-36 v2) version 2.0, World Health Organization QOL-26, and Hospital Anxiety and Depression Scale. **Results:** No significant correlation was observed between the FIM scores and scores for all three scales ($R < |0.2$ | for every item). **Conclusions:** ADLs did not correlate significantly with psychological distress or QOL without considering independent demographic characteristic factors

Keywords: Activities of daily living, Duchenne muscular dystrophy, psychological distress, quality of life

INTRODUCTION

Duchenne muscular dystrophy (DMD) is a rare X-linked disease.^[1] Advances in treatment have increased the life expectancy. The skill levels of activities of daily living (ADLs) in adults with DMD have not been exclusively studied. There seems to be no agreement whether depression and anxiety are present in adolescents and adults with DMD.^[2,3] A multinational study measured health-related quality of life (HRQOL) in the DMD population;^[4] however, QOL measures in Japanese adults with DMD are unknown. This study aimed to investigate the current state of Japanese adults with DMD and analyze the relationship of ADLs with QOL and psychological state.

PATIENTS AND METHODS

The study was approved by the Institutional Review Board of National Center of Neurology and Psychiatry, National Center Hospital of Neurology and Psychiatry that specializes in the treatment of neuromuscular diseases (A2015-095). The data

for this study were collected between March 2016 and March 2018. All study procedures were conducted according to the principles of the World Medical Association Declaration of Helsinki.

Study design and patients

This was a cross-sectional observational study.

Participants were male patients with DMD over 20 years of age who were referred to our department by neurologists or pediatric neurologists at the hospital. Inclusion criteria were the ability to provide conscious consent for participation and no developmental and/or intellectual disorders that could be considered a confounding factor.

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Table 1: Demographic characteristics of participants (n=36)

	<i>n</i> (%)	Average	Median±SD	Range
Age	36 (100)	26.3	25.0±4.7	20-36
Age at diagnosis	36 (100)	4.3	4.5±2.1	0-10
Duration of the disease (years)	36 (100)	22.0	21.5±5.0	14-30
Age at loss of ambulation (years)	36 (100)	10.2	10.0±1.6	7-14
Corticosteroid treatment				
Ongoing	5 (13.9)	23.4	20±4.7	20-29
None	15 (41.7)	27.9	30±4.6	20-33
Discontinued	16 (44.4)	25.6	24±4.6	20-36
Heart medication	36 (100)			
Operation history of scoliosis correction	4 (11.1)			
Noninvasive ventilator				
User	31 (88.9)	26.6	25.0±4.6	20-36
Initiation age (years)		19.8	19±4.0	12-29
Nonuser	5 (13.9)	24.2	20.0±5.85	20-32
The using timing of NIV (n=31)				
Only at night	13 (41.9)	24.6	24.5±3.5	20-32
All day	13 (41.9)	28.2	30.0±3.9	21-32
At night and in daytime appropriately	5 (16.1)	26.6	23.0±7.37	20-36
Nutrition				
Oral	31 (86.1)	25.9	25±4.90	20-36
With gastrostoma	4 (11.1)	29	28.0±1.73	28-31
With percutaneous transesophageal gastrotubing	1 (2.8)	31		
Wheelchair mobility				
Independent power wheelchair driving	26 (72.2)	27.1	28.5±4.76	20-36
Not using or dependent wheelchair propulsion at home.	4 (11.1)	23.75	25±2.5	20-25
Independent power wheelchair driving outside				
Not using or dependent wheelchair propulsion in and out	6 (16.7)	24	22±4.94	20-32
Upper limb function				
Possible to write by hand	13 (36.1)	23.5	22±4.1	20-32
Impossible to write by hand	23 (63.9)	27.8	29±4.4	20-36
Living status				
Alone	3 (8.3)	30.3	31±1.15	29-31
With parents	10 (27.8)	29.7	31.5±4.92	20-36
With parents and other family members	14 (38.9)	25.1	24.5±4.05	20-33
With mother	4 (11.1)	22.8	23±2.22	20-25
With one parent and other family members	5 (13.9)	23	21±3.39	20-28
Educational background				
High school graduate	10 (27.8)	25	24±4.64	20-33
Technical school graduate	1 (2.8)	36		
In college	6 (16.7)	20.5	20±0.84	20-22
College graduate	13 (36.1)	28.9	30±3.30	22-32
Incomplete college degree	3 (8.3)	26	25±4.58	22-31
Graduate school graduate	3 (8.3)	27.3	28±2.08	25-29
Employment status				
Working	14 (38.9)	27.8	29±4.02	20-32
Full time	6 (16.7)	28.8	29±2.79	24-32
Part time	8 (22.2)	27.1	27.5±4.63	20-32
Not working	22 (61.1)	25.1	23.5±5.01	20-36
Public welfare services				
Home health nurse	26 (72.2)	27	28±4.74	20-36
Home caregiver	27 (75.0)	27.4	29±4.62	20-36
Home-visit rehabilitation	22 (61.1)	26.2	26.5±4.82	20-36
Outpatient rehabilitation	29 (80.6)	26.2	25±4.73	20-33
Both home-visit and outpatient rehabilitation	15 (41.7)	26.1	28±4.85	20-32

NIV: Noninvasive positive pressure ventilation, SD: Standard deviation

Data collection

Demographic characteristics of study participants were obtained from medical records. Collected information included age, age at the time of diagnosis, duration of disease, age at loss of ambulation, management of steroids, cardiovascular medication, status of respirator use, feeding pathway, history of surgical treatment for scoliosis, educational background, and living situation [Table 1].

In one earlier study,^[5] the Functional Independence Measure (FIM) was used to assess patients' ADLs.^[6-9] To examine the emotional state of patients, the Japanese version of the Hospital Anxiety and Depression Scale (HADS) has been used.^[10] In several previous studies,^[3,11] the 36-item Short-Form Health Survey (SF-36)^[12] and the World Health Organization QOL Scale (WHOQOL-BREF)^[13] were used to assess QOL. These studies were based on an opinion expressed by Huang *et al.*,^[14] that the SF-36 could be used to measure the health-related perceived status, capabilities, and functioning as an HRQOL measurement and that the WHOQOL-BREF could be used to assess the self-satisfaction and a global QOL measurement. To calculate the overall domain score for WHOQOL-BREF, the raw item scores for each domain were used. A transformed score with the score range 4–20 was obtained by converting the domain score. This method of converting the raw scores to a transformed score was demonstrated by the WHOQOL group.^[15] Item 1 measured overall QOL and Item 2 measured general health (GH). In this study, the Japanese versions of the 36-Item Short-Form Health Survey version 2.0 (SF-36 v2)^[16] and WHOQOL-BREF (WHOQOL26)^[17] were used to assess HRQOL and global QOL.

After obtaining each participant's consent, an investigator assessed each patient to determine the FIM score. If a patient was able to independently complete the questionnaires, an investigator collected the forms in 2–3 days for inpatients or at the time of the next outpatient visit. If a patient was unable to write responses, an investigator recited each question for the patient and recorded the spoken reply.

Statistical analysis

Spearman's correlation analysis was performed to measure the association among each variable of the FIM (total, motor, and cognitive), QOL (the item scores of SF-36 v2 and the domain scores of WHOQOL26), and HADS (anxiety for HADS-A and depression for Hospital Anxiety and Depression Scale Depression subscale [HADS-D]).

For SF-36 v2, the score values were compared to age-adjusted values of Japanese national norms using Welch's one-sample *t*-test. $P < 0.05$ was considered statistically significant. Data analysis was performed using SPSS23® (IBM, Armonk, NY, USA).

RESULTS

Patients characteristics

Figure 1 shows the patients' flow. There were 36 enrolled patients and their characteristics are presented in Table 1. The

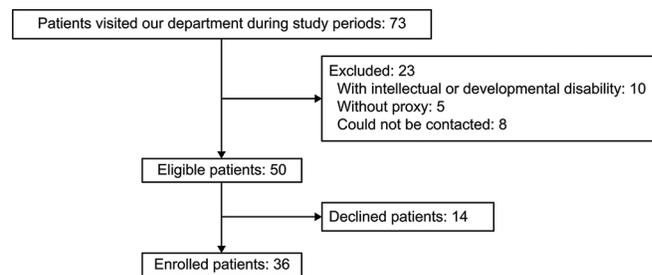


Figure 1: Patients' flow. Based on the inclusion criteria, 50 patients were eligible to participate, and 36 patients consented to participate in the study

average patient ages at the time of this study and at the time they had received the diagnosis were 26.3 ± 4.7 years (range 20–36) and 4.3 ± 2.1 years (range 0–10), respectively. Five patients with an average age of 23.4 ± 4.7 years were continuing treatment with glucocorticoids until this study period. Noninvasive positive pressure ventilation (NIV) had already been initiated in 32 patients (88.9%), with the average starting age for NIV of 20.3 ± 3.8 years old. Twenty-six patients (72.2%) were able to self-propel their electric wheelchairs. Thirteen patients (36.1%) were able to handwrite questionnaire responses. Most participants lived with family members, but three patients (8.3%) lived alone and utilized the public welfare services such as home caregivers. Fourteen patients (38.9%) were employed. Most patients were receiving either home-visit or outpatient rehabilitation service (61.1% and 80.6%, respectively).

Activities of daily livings assessed by the Functional Independence Measure

The average scores for total FIM and motor FIM were 53.9 ± 3.4 and 19.0 ± 3.1 points, respectively. The distribution of the motor scores of FIM was 13 (total assistance) to 25 (maximum assistance). Although there was no statistical significance, the scores with individual differences were in the areas of eating (2.4 ± 1.6 points), grooming (1.4 ± 0.7 points), bladder management (1.9 ± 0.4 points), bowel management (1.6 ± 0.5 points), and wheelchair locomotion (3.8 ± 1.5 points) [Table 2].

The Hospital Anxiety and Depression Scale assessment

The average scores and standard deviation for HADS-A (anxiety) and HADS-D (depression) were 4.8 ± 2.8 (maximum 14, minimum 0) points and 2.9 ± 2.5 (maximum 9, minimum 0) points, respectively. Overall, this population did not demonstrate a clinically significant state of anxiety and depression. In terms of individual differences, five patients for HADS-A and two patients for HADS-D demonstrated scores of 8–11 points, which were suggestive of anxiety or a depressive state. One patient had a HADS-A score over 11 points, indicating clinical anxiety. The probable rate of anxiety and depression was 2.8% and 0%, respectively.

The SF-36 v2 assessment

As shown in Table 3, the values of all the domains for SF-36 v2 in study participants were compared to the Japanese standard

Table 2: Distribution of Functional Independence Measure scores

	Average	Median	Maximum	Minimum	SD	Fujiwara <i>et al.</i> (2009)
Total score (126)	53.9	53	60	48	3.4	
Motor (91)	19.0	18	25	13	3.1	28.3
Cognitive (35)	34.9	35	35	30	0.8	32
Motor items						
Eating	2.4	1	5	1	1.6	
Grooming	1.4	1	4	1	0.7	
Bathing	1.0	1	1	1	0.0	
Dressing-upper	1.0	1	1	1	0.0	
Dressing-lower	1.0	1	1	1	0.0	
Toileting	1.0	1	1	1	0.0	
Bladder management	1.9	2	3	1	0.4	
Bowel management	1.6	2	2	1	0.5	
Bed/chair/wheelchair transfers	1.0	1	1	1	0.0	
Toilet transfers	1.0	1	1	1	0.0	
Bath/shower transfers	1.0	1	1	1	0.0	
Walk/wheelchair	3.8	4	5	1	1.5	
Stairs	1.0	1	1	1	0.0	
Cognitive items						
Comprehension	7.0	7	7	7	0.0	
Expression	6.9	7	7	4	0.5	
Social interaction	7.0	7	7	7	0.0	
Problem solving	6.9	7	7	5	0.3	
Memory	7.0	7	7	7	0.0	

No significant differences were found in scores for each item. Depending on the item, individual differences were found in eating (2.4 ± 1.6 points), grooming (1.4 ± 0.7 points), bladder management (1.9 ± 0.4 points), bowel management (1.6 ± 0.5 points), and wheelchair locomotion (3.8 ± 1.5 points). SD: Standard deviation

values. The physical functioning (PF) and role physical (RP) domains in study participants showed statistically significantly lower values of 7.2 ± 16.3 and 70.8 ± 25.8 ($P < 0.001$), respectively. For all other domains, there were no significant differences between study participants and the Japanese standard population.

The World Health Organization Quality of Life 26 assessment

For the WHOQOL26, the number of patients who responded “very good” or “good” was 20 (55.6%) for Item 1 and 21 (58.3%) for Item 2, respectively. The domains of psychological, social relationships, environmental, and total were higher than age-adjusted values of the Japanese national norm, with a statistically significant difference ($P < 0.001$) for the environmental domain. For study participants, the score for the physical health domain (3.33 ± 0.51) was slightly lower than the Japanese national norm (3.39 ± 0.57 , $P = 0.639$).

Correlation among the scores of the Functional Independence Measure, Hospital Anxiety and Depression Scale, SF-36 v2, and World Health Organization Quality of Life 26

As shown in Figure 2, no significant correlation was found for the FIM scores (total, motor, and cognitive) with the scores of HADS-A and HADS-D, the subscales of SF-36 v2, and the domains of the WHOQOL26. The FIM scores did not considerably affect the psychological distress, HRQOL, and global QOL. However, for the scores of the

HADS-A among SF-36 v2 and WHOQOL26, the Spearman’s correlation was detected in the subscales of GH ($\rho = -0.39$, $P < 0.05$) and social functioning (SF) ($\rho = -0.49$, $P < 0.01$) for the SF-36 v2, and the physical ($\rho = -0.343$, $P < 0.05$), psychological ($\rho = -0.584$, $P < 0.01$), social relationships ($\rho = -0.448$, $P < 0.01$), and total ($\rho = -0.402$, $P < 0.05$) domain of WHOQOL26. The HADS-D was moderately negatively correlated with SF-36 v2 domains of RP ($\rho = -0.495$, $P < 0.01$), bodily pain (BP) ($\rho = -0.516$, $P < 0.01$), GH ($\rho = -0.377$, $P < 0.05$), vitality (VT) ($\rho = -0.711$, $P < 0.01$), role emotional (RE) ($\rho = -0.451$, $P < 0.01$), and mental health (MH) ($\rho = -0.414$, $P < 0.05$). The HADS-D was also moderately negatively correlated with the WHOQOL26 domains of physical ($\rho = -0.436$, $P < 0.01$), psychological ($\rho = -0.394$, $P < 0.05$), and total ($\rho = -0.424$, $P < 0.05$). The multivariate analysis showed that independent demographic characteristic factors, such as age, glucocorticoid treatment status, initiated NIV, gastrostomy, or family structure did not correlate with the scores of FIM, HADS, SF-36 v2, or WHOQOL-26.

DISCUSSION

This study has identified physical and psychosocial features of a part of the population of Japanese adults with DMD. Skill levels of ADLs were not significantly correlated with depression, anxiety, HRQOL, and global QOL. Unlike the recent study,^[18] using the FIM scores, individual differences in

	Age	FIM			HADS		SF-36v2								WHOQOL26				
		Motor	Cognitive	Total	HADS-A	HADS-D	PF	RP	BP	GH	VT	SF	RE	MH	Physical health	Psychological	Social relationships	Environmental	Total
Age		-0.39	0.25	-0.32	-0.11	0.30	0.15	-0.07	-0.13	-0.01	-0.12	0.13	-0.13	-0.19	-0.13	-0.25	-0.05	-0.09	-0.18
FIM	Motor	-0.39	0.04	0.97	0.07	-0.11	-0.05	0.05	-0.07	-0.03	0.07	0.03	0.00	0.03	0.27	0.13	0.13	0.10	0.01
	Cognitive	0.25	0.04	0.27	0.13	0.24	-0.12	-0.03	-0.23	-0.24	0.06	0.26	-0.20	-0.04	-0.26	-0.10	-0.06	-0.17	-0.27
	Total (Motor + Cognitive)	-0.32	0.97	0.27	0.10	-0.05	-0.07	0.04	-0.12	-0.09	0.08	0.08	-0.05	0.02	0.20	0.10	0.11	0.06	-0.06
HADS	HADS-A	-0.11	0.07	0.13	0.10	0.42	-0.11	-0.27	-0.18	-0.39	-0.33	-0.49	-0.30	-0.70	-0.34	-0.58	-0.45	-0.13	-0.40
	HADS-D	0.30	-0.11	0.24	-0.05	0.42	0.13	-0.50	-0.52	-0.38	-0.71	-0.20	-0.45	-0.41	-0.44	-0.39	-0.20	-0.25	-0.42
SF-36v2	PF	0.15	-0.05	-0.12	-0.07	-0.11	0.13	0.11	-0.10	0.24	-0.05	0.25	0.00	0.10	0.22	0.10	-0.02	0.23	0.15
	RP	-0.07	0.05	-0.03	0.04	-0.27	-0.50	0.11	0.23	0.20	0.46	0.72	0.75	0.35	0.43	0.28	0.17	0.44	0.29
	BP	-0.13	-0.07	-0.23	-0.12	-0.18	-0.52	-0.10	0.23	0.34	0.54	0.05	0.18	0.39	0.20	0.22	0.11	0.25	0.47
	GH	-0.01	-0.03	-0.24	-0.09	-0.39	-0.38	0.24	0.20	0.34	0.32	0.05	0.02	0.26	0.61	0.62	0.45	0.42	0.54
	VT	-0.12	0.07	0.06	0.08	-0.33	-0.71	-0.05	0.46	0.54	0.32	0.25	0.32	0.47	0.43	0.45	0.32	0.30	0.52
	SF	0.13	0.03	0.26	0.08	-0.49	-0.20	0.25	0.72	0.05	0.05	0.25	0.56	0.46	0.28	0.29	0.13	0.33	0.23
	RE	-0.13	0.00	-0.20	-0.05	-0.30	-0.45	0.00	0.75	0.18	0.02	0.32	0.56	0.37	0.21	0.18	0.07	0.20	0.16
	MH	-0.19	0.03	-0.04	0.02	-0.70	-0.41	0.10	0.35	0.39	0.26	0.47	0.46	0.37	0.29	0.53	0.35	0.11	0.43
WHOQOL26	Physical health	-0.13	0.27	-0.26	0.20	-0.34	-0.44	0.22	0.43	0.20	0.61	0.43	0.28	0.21	0.29	0.67	0.68	0.59	0.63
	Psychological	-0.25	0.13	-0.10	0.10	-0.58	-0.39	0.10	0.28	0.22	0.62	0.45	0.29	0.18	0.53	0.67	0.70	0.34	0.66
	Social relationships	-0.05	0.13	-0.06	0.11	-0.45	-0.20	-0.02	0.17	0.11	0.45	0.32	0.13	0.07	0.35	0.68	0.70	0.24	0.47
	Environmental	-0.09	0.10	-0.17	0.06	-0.13	-0.25	0.23	0.44	0.25	0.42	0.30	0.33	0.20	0.11	0.59	0.34	0.24	0.49
	Total	-0.18	0.01	-0.27	-0.06	-0.40	-0.42	0.15	0.29	0.47	0.54	0.52	0.23	0.16	0.43	0.63	0.66	0.47	0.49

Figure 2: Spearman's correlation among the measurements. Darker color of cells shows a higher correlation among each domain. No significant differences were observed in the scores of FIM among HADS, the SF-36 v2, and the WHOQOL26. FIM: Functional independence measure, HADS-A and HADS-D: Hospital Anxiety and Depression Scale–Anxiety Subscale and Hospital Anxiety and Depression Scale–Depression measure, SF-36 v2: The 36-Item Short-Form Health Survey version 2.0, PF: Physical functioning, RP: Role physical, BP: Bodily pain, GH: General health, VT: Vitality, SF: Social functioning, RE: Role emotional, MH: Mental health, WHOQOL-BREF: World Health Organization Quality of Life Scale-BREF

Table 3: Scores for each domain of the 36-item Short-Form Health Survey and World Health Organization Quality of Life-26 compared with the Japanese norm

Domains	Average±SD		Mean difference	P
	Patients	Japanese norm		
SF-36v2				
PF	7.2±16.3	89.1±13.9	-9.14	<0.001
RP	70.8±25.8	89.2±18.8	40.21	<0.001
BP	72.1±20.9	73.8±22.4	49.26	0.641
GH	56.6±17.7	62.9±18.8	46.63	0.041
VT	64.1±20.1	62.8±19.5	50.63	0.719
SF	77.1±25.9	86.4±19.4	45.21	0.041
RE	79.2±18.5	87.8±20	45.66	0.009
MH	72.8±13.7	71.6±18.6	50.63	0.616
WHOQOL-26		(Age-adjusted)		
Physical health	3.33±0.51	3.39±0.57		0.639
Psychological	3.61±0.52	3.23±0.59		0.005
Social relationships	3.25±0.54	3.06±0.65		0.182
Environmental	3.67±0.53	3.09±0.53		<0.001
Total	3.49±0.72	3.19±0.43		0.035

For SF-36v2, scores for adults with DMD and the Japanese standard were compared. A significant difference was seen in the domains of PF, RP, GH, SF, and RE, with extremely low scores in the domains of PF and RP. For WHOQOL26, average scores for study participants were slightly higher than the Japanese standard values levels by age group, except for the physical domain. PF: Physical functioning, RP: Role physical, BP: Bodily pain, GH: General health, VT: Vitality, SF: Social functioning, RE: Role emotional, MH: Mental health, SD: Standard deviation, SF-36v2: The 36-item Short-Form Health Survey version 2, WHOQOL26: World Health Organization Quality of Life-26, DMD: Duchenne muscular dystrophy

gross and fine motor skills for adults with DMD can be roughly assessed. For the scores of total and motor FIM, even though most of the study participants needed anywhere from total assistance to maximum assistance for ADLs with significantly reduced physical domains of HRQOL, the existence of psychological distress in this population was found to be in the normal range without clinical depression and anxiety.

A previous study revealed that seven Japanese adults with DMD (average age of 37.3 years old) felt anxiety and worry when they became wheelchair and respirator dependent.^[19] For participants in the present study, none of the physical and social characteristics were found to be statistically relevant factors for depression and anxiety, even though all the patients were wheelchair dependent and 31 (88.9%) were using noninvasive ventilators. None of our patients were using tracheostomy with invasive ventilation (TIV); thus, we could not determine whether TIV influenced the development of depression and anxiety. One study revealed that nonambulatory DMD patients had a higher rate of depression than ambulatory patients.^[20] A Dutch study of adults with DMD revealed that the average symptom scores of HADS-A and HADS-D were higher, and the average age was older than the population in the present study.^[3] Furthermore, 11.4% in the Dutch study received antidepressant medication, fatigue was present in 40.5%, and 73.4% reported BP.^[3] In the UK study, the perception of pain in the DMD patient group was significantly higher than in the control group.^[18] In the present study, based on the item scores of the SF-36 v2, 75% and 27.7% of patients reported body pain and usual fatigue, respectively. Therefore, the existence of depression could be suggested as causes for the difference in the prevalence of psychological distress between the Dutch and current studies.

In the SF-36 v2 results for the current study, the PF and RP domain scores were significantly lower than the national standard value of Japanese which was similar to results obtained from several other studies.^[3,11,18,21,22] In Germans with DMD over the age of 18, all the domains of physical scales (PF, RP, BP, and GH) were significantly lower, but SF, RE, and MH in psychological scales showed no difference between study participants and healthy German subjects.^[21] In the previously mentioned Dutch study, adults with DMD were compared to a Dutch reference group, the domain of RE was similar in both groups, with a higher score in PF, lower

scores in other domains in the patient's group, and BP and VT scores not significantly lower.^[3] In the current study results, PF and RP were significantly low; however, domain scores for VT and MH were slightly higher than the Japanese standard norm. These findings were different from the UK study in which all of the domain scores were lower than control subjects.^[18] The German study included three patients who were able to walk <50 m, nocturnal ventilation was used in 50% of the subgroup, and 20.8% had a gastrostoma.^[21] The Dutch group included participants with severe complication, including 53.2% who needed TIV, 45.6% who needed NIV, and 27.9% with gastrostoma.^[3] In addition, 34.1% of participants had already lived in a residential home.^[3] From these findings, the different distribution trends in SF-36 domains between the current and previous studies may be caused by the functional severity, seriousness of complications, and different social background among the patients in each study. Interestingly, the current results of WHOQOL26 showed not significant but slightly lower scores of physical health, while the other domain scores were slightly higher than the Japanese national age-adjusted norm with the significance in the environmental domain. In the WHOQOL-BREF result of Taiwanese DMD patients, moderate-to-large impact was noted in the domain of physical health and psychological and social relationships with the significant difference in the domain score of social relationships between younger and older DMD patients.^[11] Furthermore, only significant difference was found in social relationships domain in Dutch adult DMD patients compared with the standard norm.^[3] When comparing the current results of the SF-36 v2 and the WHOQOL26 to those of previous studies, the domains of PF and physical health were lower than the national standard norm, which was similar to the earlier studies. However, trends in the distribution of other domain scores were unequal to the other countries, perhaps due to the perception that global QOL may also be affected by their functional complication, surrounding culture, and the living situation of the study participants. Similar to the previous study,^[3] the PF and RF domains of HRQOL in our patients were markedly affected by their disease; however, their global QOL seems to maintain as much as similar degree to age-adjusted healthy Japanese subjects.

In this study, to clarify whether the skill levels of ADLs may have a potential impact among psychological distress, HRQOL, and global QOL, we examined the significance of correlation among the FIM, HADS, SF-36 v2, and WHOQOL26. However, no conspicuous correlation was found similar to the recent study.^[18] A study in Taiwanese individuals with DMD showed that the basic ADLs score had a moderate correlation among the domains of PF and SF on the SF-36 v2, and correlated between the domains of social relationships of the WHOQOL-BREF.^[11] This differed from the results of the current study. A possible cause for this difference was the average age in the current study, which was higher than the study group.^[11] In addition, the current study used the FIM for the evaluation of ADLs, which was not a specific measure for

MD. It differed from the study where they used one domain from the scale that they designed particularly for patients with MD. Fujiwara *et al*. reported that the mean FIM motor score for Japanese individuals with DMD over 15 years old was 28.3 (ranging from 14 to approximately 50),^[23] while our result was 19.0 with very low variation (ranging from 13 to 25). This means that the distribution of the motor FIM scores of participants in the current study is not varied [Table 2]. Correlation between factors is difficult to detect in data with low variation, such as FIM score in the current study. An Italian study assessed 48 ventilated adults with DMD using the individualized neuromuscular QOL, which was developed as a specialized measurement tool to assess HRQOL in patients with muscular diseases. Their results showed poor QOL, with the weakest domains being physical health and area of life, while the psychosocial domain was not as weak.^[24] Even though not all participants in our study used NIV, the distribution tendency of the HRQOL domain obtained nearly the same results as that of the Italian study. In addition, Otto *et al*. showed that HRQOL was not significantly associated with the age of disease onset.^[25] Although people with serious health conditions with persistent disabilities and impairments can experience good QOL, Albrecht and Devlieger described these individuals with the phrase "disability paradox."^[26] Disability paradox in individuals with DMD was mentioned by Pangalila^[27] and seen in a multinational study.^[4] It has been described that patients in the early stage of DMD show a "response shift," wherein individuals who confront such health changes, the criteria for judging answers for QOL or patient-reported outcome measures are changing.^[28] This phenomenon may enable them to experience high QOL.^[27]

Even our study detected the disability paradox of psychological disability and HRQOL in study participants as well as in a precedence study.

Some limitations exist in the study. First, the personality traits of patients may present some bias. Consented patients may be more curious and motivated to try unfamiliar activities. Second, to use the validated questionnaires to investigate mental status and QOL, patients were excluded who were unable to comprehend the contents and answer the questions. Furthermore, the examiner asking the patients the questions and their responses being recorded by the examiner might have influenced the answers. Third, the gene abnormalities of each patient were not considered in this study. It is reported that part of the mutation affects the neuropsychological recognition of a patient's traits, psychological status, and recognition for their QOL.^[29] Further study is needed to evaluate methods other than questionnaires to assess psychosocial factors impacting adults with DMD and to examine the types and parts of gene mutations that may affect the perception of psychological distress and QOL. This study demonstrated physical, social, and psychological characteristics of a part of the population of Japanese adults with DMD. Even if our patients' ADLs levels were almost dependent on the others, our results show that their mental status and QOL could be maintained in favorable

conditions without severe perception of anxiety, depression, and pain. Since DMD is an intractable rare disease, there are not many studies focusing only on adults.^[3,18] The results of the current study provide meaningful information for Japanese adults with DMD and their caregivers. Moreover, physicians should consider not only maintaining physical functions of patients but also promoting social participation in consideration of the patients' respiratory and cardiac conditions.

CONCLUSION

This study investigated the physical and psychosocial characteristics of a part of the Japanese population of adults with DMD. Almost all study participants required assistance with ADLs, with slight individual differences. The skill levels of ADLs had no statistically significant correlation with anxiety, depression, HRQOL, and global QOL. Furthermore, no significant correlations were found for the physical and social characteristics of patients among psychological distress and QOL.

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Conflicts of interest

There are no conflicts of interest.

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