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Comparison Between Courses of Home and Inpatients Mechanical Ventilation in Patients with Muscular Dystrophy in Japan

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1. Introduction

In Japan, 27 hospitals specialize in treatment of muscular dystrophy patients, including inpatient care, of which 26 belong to the National Hospital Organization, and the other is the National Center of Neurology and Psychiatry. Since 1999, Japanese muscular dystrophy research groups investigating nervous and mental disorder have been developing a database of cases treated at these 27 institutions. In that regard, we conducted a survey of inpatients and home-mechanical ventilation patients (HMV patients) with muscular dystrophy and other neuromuscular disorders based on data collected by the National Hospital Organization and National Center of Neurology and Psychiatry.

Herein, we examined data obtained in order to evaluate efficacy of mechanical ventilation therapy for HMV patients and mechanical ventilation-dependent inpatients (MV inpatients) with those wards.

2. Subjects and methods

The database includes numbers of inpatients, gender, age, diagnosis, respiratory condition, nutritional state, number of death cases, causes of death, and other relevant findings from data collected annually on October 1 every year since 1999. Additionally we collected the data of HMV patients from 27 institutes for this study.

By using the database and newly collected HMV data, we analyzed the courses of HMV patients group and those of MV inpatients of wards. We compared data of these two groups. Examination points are mechanical ventilation periods, outcome of these two groups, and caregiver for HMV patients.

2.1 Objective diseases

Objective diseases of this study were muscular dystrophy and spinal muscular atrophy, in particular Duchenne muscular dystrophy and myotonic dystrophy. Amyotrophic lateral sclerosis was not included.

2.2 Patients introduced HMV after 1999

The data which we requested 27 institutes specializing muscular dystrophy care was as follows; the number of patients introduced HMV after 1999, diagnosis of disease, gender, age at being introduced HMV, type of mechanical ventilation, such as non-invasive positive pressure ventilation (NPPV) or tracheostomy intermittent ventilation (TIV), present status, death cause for death case, main caregiver, and so on.

2.3 Patients introduced MV in muscular dystrophy wards after 1999

We selected data of newly MV introduced inpatients after 1999 from the database of the muscular dystrophy wards.

3. Results

3.1 Demographic features of HMV patients group and MV inpatients

3.1.1 HMV patients group

HMV patients group included 434 patients from 14 institutes. Gender was male: 356, female: 78. The number of representative disease were as follows; 262 patients with Duchenne muscular dystrophy, 60 myotonic dystrophy, 17 Becker muscular dystrophy, 16 limb-girdle muscular dystrophy, 14 spinal muscular atrophy, and so on (Table 1-1).

Diagnosis	HMV	Inpatient	Total	Death cases
BMD	17	35	52	10
CMD	12	6	18	2
DMD	262	476	738	96
EDMD	2	0	2	
FCMD	16	43	59	13
FSHD	6	33	39	7
LGMD	16	42	58	12
MD	60	222	282	62
MG	1	0	1	
SMA	13	19	32	6
SPMA	0	11	11	4
UCMD	9	1	10	
Mitochondria disease	0	5	5	1
Distal myopathy	2	3	5	2
Congenital myopathy	8	12	20	
Glycogen storage disease	2	1	3	
Other myopathies	3	1	4	
Other dystrophies	4	5	9	
Unknown	1	0	1	
total	434	915	1349	215

BMD, Becker muscular dystrophy; CMD, congenital muscular dystrophy; DMD, Duchenne muscular dystrophy; EDMD, Emery-Dreifuss muscular dystrophy; FCMD, Fukuyama congenital muscular dystrophy; FSHD, facio-scapulo-humeral muscular dystrophy; LGMD, limb-girdle muscular dystrophy; MD, Myotonic dystrophy; MG, myasthenia gravis; SMA, spinal muscular atrophy; SPMA, spinal progressive muscular atrophy, UCMD, Ullrich congenital muscular dystrophy

Table 1-1. Details of disease (HMV: from 14 institutes)

3.1.2 MV inpatients group

MV inpatients group included 915 inpatients. Gender was male: 718, female: 197. The number of representative disease were as follows; 476 Duchenne muscular dystrophy, 222 myotonic dystrophy, 35 Becker muscular dystrophy, 58 limb-girdle muscular dystrophy, 19 spinal muscular atrophy, and so on (Table 1-1).

3.1.3 Mean age at starting mechanical ventilation and type of ventilation

The range of mechanical ventilation introduction age for HMV patients was 6.3~72.8 years old (mean 25.9), and that of MV inpatients was 0.0~78.0 years old (mean 33.2). The number of NPPV introduction cases of HMV patients was 420, and that of MV inpatients was 517 (Table 1-2). Fifty of NPPV cases of HMV group were switched to tracheostomy during the course.

	HMV	Inpatient	
Age at starting mechanical ventilation (years old)	25.9	33.2	p<0.05
NPPV introduced case	420	517	

Table 1-2. Mean age at starting mechanical ventilation and type of ventilation

3.2 Survival analysis of HMV patients group and MV inpatients

We performed survival analysis of those two groups. The endpoint for HMV patients was death or transition to hospitalization, and that for MV inpatient was death. Kaplan-Meier analysis showed that 75% life time of HMV patients was 1,689 days, while that of inpatients was 2,988 days (Log Rank (Mantel-Cox) p<0.01) (Fig. 1).

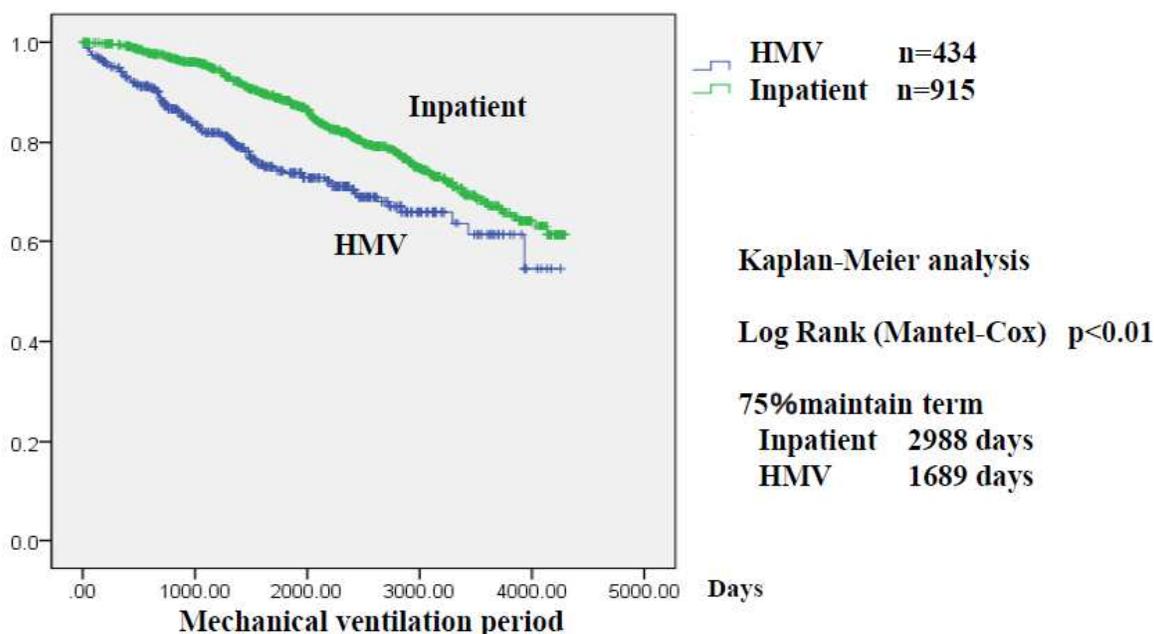


Fig. 1. Comparison between HMV Patients and Mechanical Ventilation Inpatients (Total)
Endpoint for HMV patient: death or transition to hospitalization
Endpoint for MV inpatient: death

3.3 Analysis data of Duchenne muscular dystrophy and myotonic dystrophy

As the number of patients with Duchenne muscular dystrophy and myotonic dystrophy was great in these two groups, we analyzed the data of Duchenne muscular dystrophy and myotonic dystrophy separately.

3.3.1 Mean age at starting mechanical ventilation and type of ventilation of patients with Duchenne muscular dystrophy

Mean age at starting mechanical ventilation of Duchenne muscular dystrophy was 19.8 years old, ranged from 11.5 to 39.9 years old. While that of inpatient with Duchenne muscular dystrophy was 21.5 years old, ranged from 10.0 to 42.0 years old. There was no significant difference. The number of NPPV introduction cases of HMV patients with Duchenne muscular dystrophy was 220, and that of MV inpatients was 338 (Table 2).

	HMV	Inpatient	
Age at starting mechanical ventilation (years old)	19.8	21.5	NS
NPPV	220	338	
TIV	42	138	
total	262	476	

Table 2. Mean age at starting mechanical ventilation and type of mechanical ventilation (Duchenne muscular dystrophy)

3.3.2 Type of nutrition of patients with Duchenne muscular dystrophy

The number of patients who required tube feeding, including a nasal or oral nutrition tube, and undergoing a percutaneous endoscopic gastrostomy (PEG) was apparently greater in MV inpatients group than HMV group (Table 3).

	HMV	Inpatient
Oral nutritional supply	118	314
PEG	11	78
Tube feeding (per nasal or per oral)	3	71
Intravenous hyperamelaition	0	13
unknown	130	0
total	262	476

Table 3. Type of nutrition (Duchenne muscular dystrophy)

3.3.3 Survival analysis of two Duchenne muscular dystrophy groups

We performed survival analysis of those two Duchenne muscular dystrophy groups. The endpoint for HMV patients was death or transition to hospitalization, and that for MV inpatient was death. Kaplan-Meier analysis showed that 75% life time of HMV patients was 1,562 days, while that of inpatients was 3,739 days (Log Rank (Mantel-Cox) $p < 0.01$) (Fig. 2).

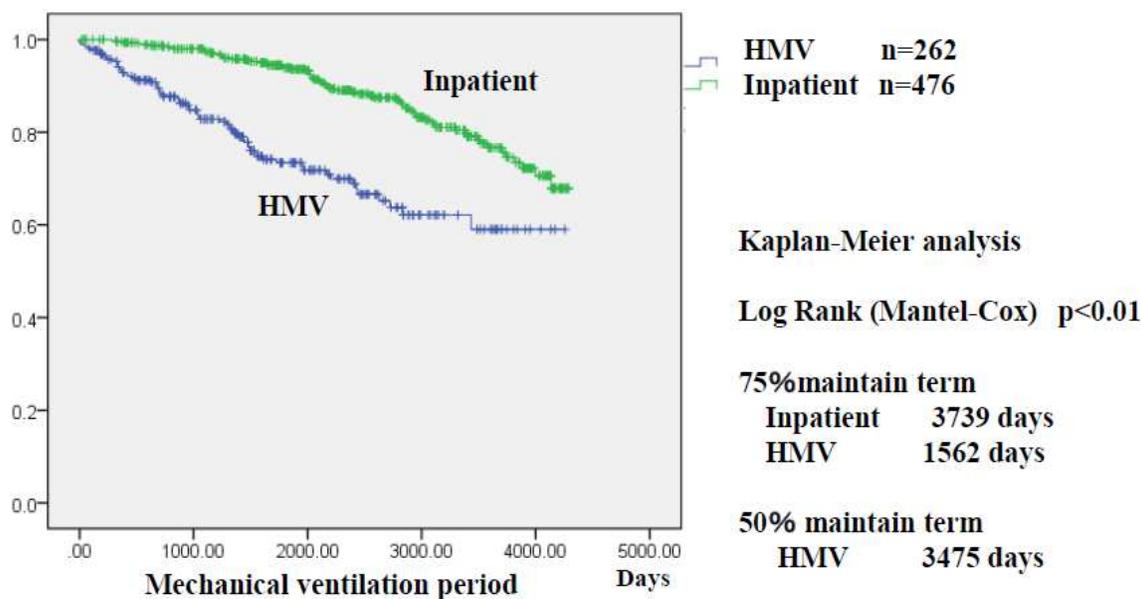


Fig. 2. Comparison between HMV Patients and Mechanical Ventilation Inpatients (Duchenne muscular dystrophy)
 Endpoint for HMV patient: death or transition to hospitalization
 Endpoint for MV inpatient: death

3.3.4 Mean age at starting mechanical ventilation and type of ventilation of patients with myotonic dystrophy

Mean age at starting mechanical ventilation of myotonic dystrophy was 46.8 years old, ranged from 15.8 to 72.8 years old. While that of inpatient with myotonic dystrophy was 50.6 years old, ranged from 12.0 to 76.0 years old. There was no significant difference between two groups. The number of NPPV introduction cases of HMV patients with myotonic dystrophy was 55, and that of TIV was 5. The number of NPPV case was greater than TIV. While the number of NPPV introduction cases of MV patients with myotonic dystrophy was 108, and that of TIV was 114. In MV patients with myotonic dystrophy, the number and proportion of NPPV and TIV were almost equal (Table 4).

	HMV	Inpatient	
Age at starting mechanical ventilation (years old)	46.8	50.6	NS
NPPV	55	108	
TIV	5	114	
total	60	222	

Table 4. Mean age at starting mechanical ventilation and type of mechanical ventilation (myotonic dystrophy)

3.3.5 Type of nutrition of patients with myotonic dystrophy

The trend of nutrition was similar to Duchenne muscular dystrophy. The number who required tube feeding in MV inpatients group was apparently greater than HMV group (Table 5).

	HMV	Inpatient
Oral nutritional supply	18	93
PEG	1	69
Tube feeding (per nasal or per oral)	0	50
Intravenous hyperamelaition	0	10
unknown	41	0
total	60	222

Table 5. Type of nutrition (myotonic dystrophy)

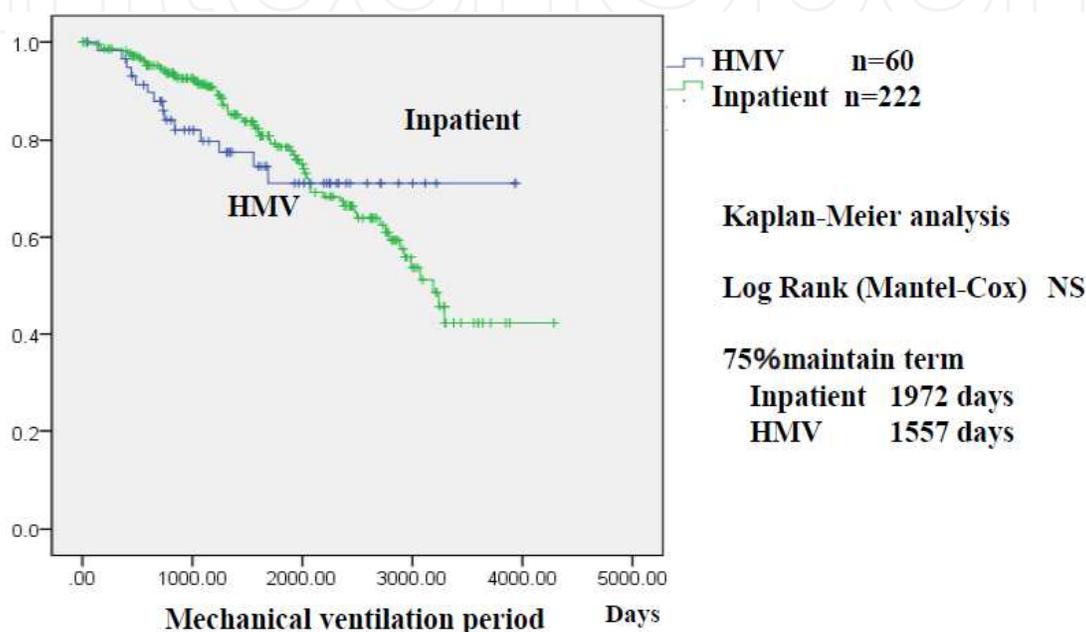


Fig. 3. Comparison between HMV Patients and Mechanical Ventilation Inpatients (myotonic dystrophy)

Endpoint for HMV patient: death or transition to hospitalization

Endpoint for MV inpatient: death

3.3.6 Survival analysis of two myotonic dystrophy groups

Similarly, we performed survival analysis of those two myotonic dystrophy groups. The endpoint for HMV patients was death or transition to hospitalization, and that for MV inpatient was death. Kaplan-Meier analysis showed that 75% life time of HMV patients was 1,557 days, while that of inpatients was 1,972 days. There was no significance (Fig. 3).

3.4 Death case

The total number of death cases was 215 (Table 1-1). As the number of death case of Duchenne muscular dystrophy and myotonic dystrophy was the majority, we analyzed the data of Duchenne muscular dystrophy and myotonic dystrophy separately.

3.4.1 Cause of death of Duchenne muscular dystrophy

The number of death cases of HMV patients was 29, and that of MV inpatients was 67.

The most frequent cause of death was heart related disorders, such as heart failure and arrhythmia, accounting for 16/29 for HMV group and 33/67 for MV inpatient group. Frequency was not significantly different between two groups. In MV inpatient group, respiratory related disorders, such as respiratory failure and respiratory infection, accounted for 23/67. HMV group included more sudden death cases than MV inpatient, and had an accidental case (Table 6).

	HMV	Inpatient	Total
Heart failure	13	27	40
Arrhythmia	3	6	9
Respiratory failure	2	11	13
Respiratory infection	1	12	13
Tracheal bleeding	1	0	1
Pneumothorax	0	1	1
Renal failure	0	1	1
Infectious disease	2	0	2
Malignancy	1	0	1
Ileus	1	0	1
Sudden death	4	2	6
Power supply accident	1	0	1
Others	0	7	7
total	29	67	96

Table 6. Cause of death (Duchenne muscular dystrophy)

3.4.2 Cause of death of myotonic dystrophy

The majority of death case of myotonic dystrophy was reported from MV inpatient group. The most frequent cause was respiratory related disorders, such as respiratory tract infection and respiratory failure, which accounted for 29/56. Sudden death case was conspicuous in HMV group, accounting for 3/6 (Table 7).

	HMV	Inpatient	Total
Respiratory infection	1	19	20
Respiratory failure	2	10	12
Heart failure	0	8	8
DIC	0	1	1
MOF	0	2	2
Cholangitis	0	2	2
Ileus	0	1	1
Choking	0	1	1
Malignancy	0	2	2
Intestinal bleeding	0	1	1
Hepatic failure	0	1	1
Sudden death	3	2	5
Others	0	6	6
total	6	56	62

Table 7. Cause of death (myotonic dystrophy)

3.5 Outcome of HMV patients and MV inpatients with Duchenne muscular dystrophy and myotonic dystrophy

One hundred ninety four cases with Duchenne muscular dystrophy among 262 cases continued HMV, while 46 cases with myotonic dystrophy among 60 cases continued HMV (Table 8). Twenty two cases with Duchenne muscular dystrophy were switched to hospitalization.

	DMD		MD	
	HMV	Inpatient	HMV	Inpatient
Continuing HMV	194	-	46	-
Continuing hospitalization	-	407	-	151
Death	29	67	6	56
Transition to hospitalization	22	-	3	-
Introduction to other institution	10	-	-	-
Withdrawing MV	-	-	3	-
Unknown	7	2	-	-
Others	-	-	2	15
total	262	476	60	222

Table 8. Outcome (Duchenne muscular dystrophy and myotonic dystrophy)

3.6 Caregivers for HMV patients

Caregivers for most of HMV patients with Duchenne muscular dystrophy were patients' families (Table 9). In particular, patients' mothers were playing important role in continuing HMV. Similarly, caregivers for HMV patients with myotonic dystrophy were patients' families. On reflecting their age, some caregivers were patient' spouse (Table 9).

Caregiver	DMD	MD
Mother	94	6
Parents	22	
Father	3	2
Family	3	
Mother/sibling	1	
Mother/uncle	1	
Mother/grandmother	1	
Grandmother	1	
Husband		2
Wife		1
Sister		1
daughter		1
foundation		2
Helper	2	2
(Response)	(134/262)	(17/60)

Table 9. HMV-continuing cases main caregiver (Duchenne muscular dystrophy and myotonic dystrophy)

3.7 Summary of results of Duchenne muscular dystrophy and myotonic dystrophy

Proportion of TIV was higher in MV inpatients group than HMV group. And proportion of tube feeding was also higher in MV inpatients group than HMV group. Namely, respiratory condition and nutritional status were more severe in MV inpatients group than HMV group (both Duchenne muscular dystrophy and myotonic dystrophy).

In Survival analysis, outcome of patients with Duchenne muscular dystrophy of MV inpatients group was better than HMV group. Meanwhile in that of myotonic dystrophy, HMV group was better than inpatient group.

In MV inpatient group of Duchenne muscular dystrophy, respiratory related death was remarkable.

In HMV group, some sudden death cases and accidental death case were conspicuous.

Caregivers of HMV group were constructed by patients' families, centrally mother.

4. Conclusion

Approximate 2500 beds for patients with muscular dystrophy and related disorders are now provided among 27 institutions in Japan. In accordance with progress in therapeutic strategies for respiratory failure (American Thoracic Society Documents, 2004) and heart failure (Ishikawa, 1999; Matsumura, 2010), the life span of patients with muscular dystrophy prolonged (Bushby 2010a, b). Now, most inpatients admitted to muscular dystrophy wards have severe general conditions and many are assisted by mechanical ventilation (Tatara, 2006; 2008), which is accordance with our data of MV patients in this study.

In recent two decades, social welfare systems and home medical care systems in Japan have been changing gradually. HMV has been penetrating into home medical care (Joseph, 2007). The number of HMV patients has been increasing (Tatara, 2006). Stable mechanical ventilated patients have been getting back home.

Our study demonstrated that the course of HMV patients was fairly good, although there was difference between Duchenne muscular dystrophy and myotonic dystrophy in long term outcome. However, the support system for patients and caregivers is not perfect. Our study also showed that burden of caregivers was supposed to be severe. The system for patients and caregivers should be adjusted (Dybwik, 2011). And safety net systems also should be adjusted to avoid accidental event leading to patient's death.

The muscular dystrophy wards may be requested to offer the circumstances for those who have difficulties in continuing HMV. There is necessarily needs for hospitalization of HMV patients (Windisch, 2008).

Study limitation: This study has limitation on bias of collecting patients' information. Specifically, information of HMV patients were reported from 14 institutes among 27 institutes, and MV inpatient information is the result of extraction from muscular dystrophy wards database. Extracted data from database has some ambiguous points in connection with obscure time-sequential analysis.

On analysis of institutes-restricted HMV patients group and MV inpatients group, differences in regard to therapeutic conditions among institutions may be problem.

5. Acknowledgments

This study was supported by a Research Grant for Nervous and Mental Disorders from the Ministry of Health, Labour and Welfare of Japan.

We are grateful to the members of the FUKUNAGA (1999-2005) and SHINNO (2006-2011) muscular dystrophy research groups of the National Hospital Organization for the data collection.

Institutions specializing in muscular dystrophy treatment in Japan (Fig.4)

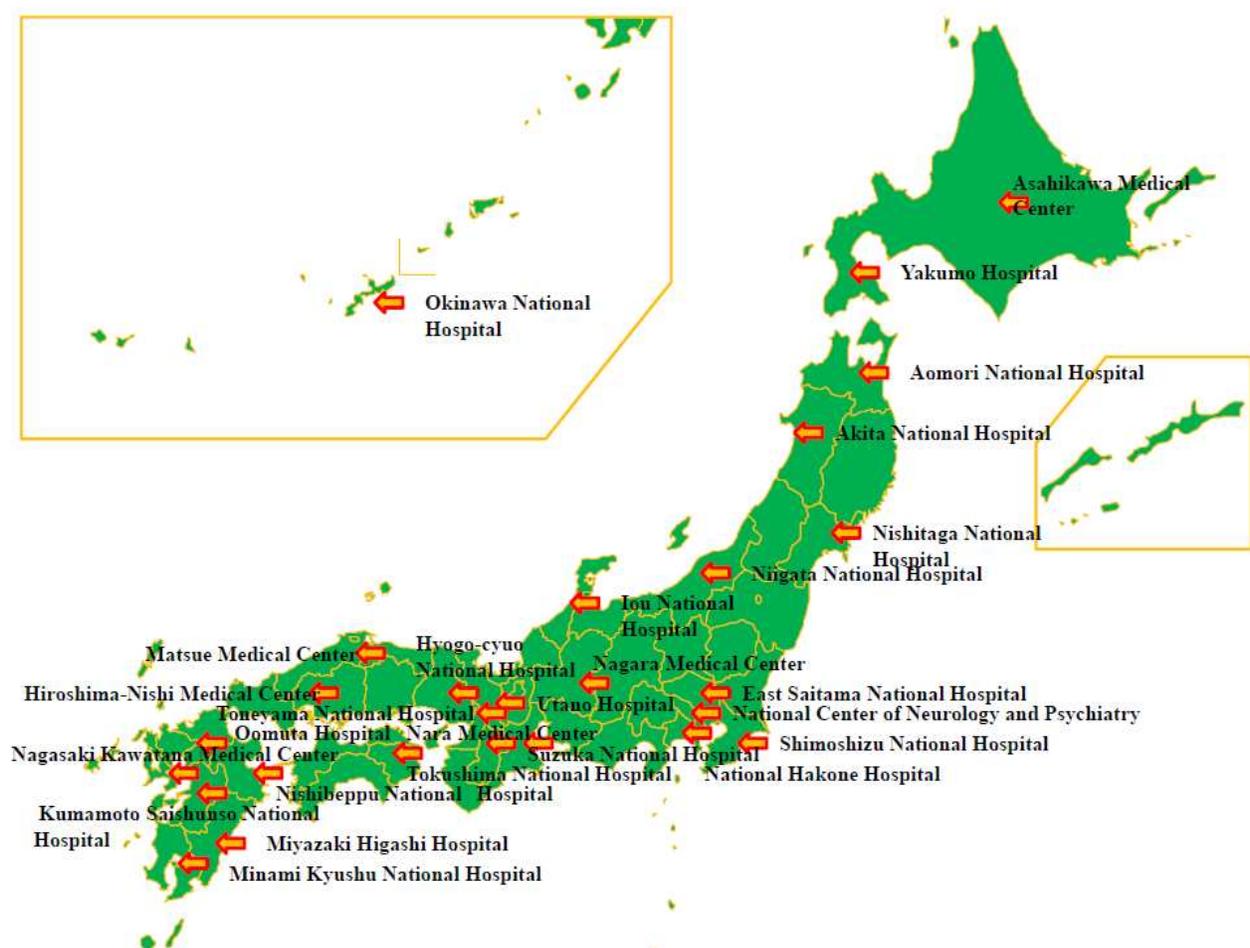


Fig. 4. Institutions specializing in muscular dystrophy treatment in Japan

National Hospital Organization:

Asahikawa Medical Center, Yakumo Hospital, Aomori National Hospital,
Akita National Hospital, Nishitaga National Hospital, East Saitama National Hospital,
Shimoshizu National Hospital, National Hakone Hospital, Niigata National Hospital,
Iou National Hospital, Nagara Medical Center, Suzuka National Hospital,
Nara Medical Center, Utano Hospital, Toneyama National Hospital,
Hyogo-cyuo National Hospital, Hiroshima-Nishi Medical Center, Matsue Medical Center,
Tokushima National Hospital, Oomuta Hospital, Nagasaki Kawatana Medical Center,
Kumamoto Saishunso National Hospital, Nishibeppu National Hospital,
Miyazaki Higashi Hospital, Minami Kyushu National Hospital, Okinawa National Hospital

National Center of Neurology and Psychiatry

6. References

- American Thoracic Society Documents (2004). Respiratory Care of the Patient with Duchenne Muscular Dystrophy. ATS Consensus Statement *American Journal of Respiratory and Critical Care Medicine*, Vol 170, pp 456-465.
- Bushby K, Finkel R, Birnkrant DJ, Case LE, Clemens PR, Cripe L, Kaul A, Kinnett K, McDonald C, Pandya S, Poysky J, Shapiro F, Tomezsko J, Constantin C, for the DMD care considerations working group (2010a). Diagnosis and management of Duchenne muscular dystrophy, part 1: diagnosis, and pharmacological and psychosocial management. *The Lancet Neurology*, Vol.9, pp 77-93.
- Bushby K, Finkel R, Birnkrant DJ, Case LE, Clemens PR, Cripe L, Kaul A, Kinnett K, McDonald C, Pandya S, Poysky J, Shapiro F, Tomezsko J, Constantin C, for the DMD care considerations working group (2010b). Diagnosis and management of Duchenne muscular dystrophy, part 2: implementation of multidisciplinary care. *The Lancet Neurology*, Vol.9, pp 177-189.
- Dybwik.K, Nielsen.E.W,Brinchmann. B. S (2011).Home mechanical ventilation and specialized health care in the community: Between a rock and a hard place. *BMC Health Services Research*, Vol. 11, pp 115-123.
- Ishikawa Y, Bach JR, Minami R (1999). Cardioprotection for Duchenne's muscular dystrophy. *American Heart Journal* Vol.137, pp 895-902.
- Joseph S. L, Peter C. G (2007). Current Issues in Home Mechanical Ventilation. *Chest*, Vol. 132, pp 671-676.
- Matsumura T, Tamura T, Kuru S, Kikuchi Y, Kawai M (2010) · Carvedilol can prevent cardiac events in Duchenne muscular dystrophy. *Internal Medicine* Vol. 49, pp 1357-1363.
- Tatara K, Fukunaga H, Kawai M (2006). Clinical survey of muscular dystrophy in hospitals of National Hospital Organization. *IRYO*, Vol. 60, pp 112-118.

Tatara K, Shinno S (2008). Management of mechanical ventilation and prognosis in Duchenne muscular dystrophy. *IRYO*, Vol. 62, pp 566-571.

Windisch W; Quality of life in home mechanical ventilation study group (2008). Impact of home mechanical ventilation on health-related quality of life. *Eur Respir J*. Vol. 32, pp 1328-1336.

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