**評価用紙・報告書の記載時の注意点**

評価用紙(別紙 1)および報告書(別紙 2)は各々の記入例を参考に作成する。

**最終日カンファレンス時の指導する際の注意点**

カンファレンスでは、多職種スタッフに対し、患者が現在服用中の薬剤について用法用量、問題点などを説明する。過去に入院歴がある場合は、前回入院時と比べて変更があった薬剤についても報告する。また多職種からの情報や意見をもとに検討する事項がある場合は検討をする。

また患者に対して、現在服用中の薬剤について、患者または薬剤の管理者に対し、再度用法用量、効果、副作用、日常生活での注意点などを説明する。また、多職種で検討した内容で患者に説明が必要な事項がある場合は説明をする。

（別紙1）

**評価用紙**　　患者名＿＿＿＿＿＿＿＿＿＿＿＿＿＿様　　回答者＿＿＿＿＿＿＿＿＿＿＿＿＿＿

| 項目 | 質問 | 回答 | コメント |
|---|---|---|---|
| 薬の管理者 | 薬は誰が管理しているか？ | □本人　□家族<br>□その他（　　） | |
| 薬の理解度 | 薬効は？ | □良好　□やや良好　□半分ほど<br>□やや不良　□不良 | |
| | 用法は？ | □良好　□やや良好　□半分ほど<br>□やや不良　□不良 | |
| 投与法 | 剤形は？ | □錠剤のみ　□錠剤＋粉薬<br>□粉薬（液剤）のみ　□内服なし | |
| | 投与経路は？ | □経口　□経鼻<br>□胃婁　□その他 | |
| | 薬の服用に介助を必要としているか？ | □不要　□必要 | |
| | 投与法にこだわりはあるか？ | □なし　□あり | |
| 飲みにくさ | 飲こみにくさはあるか？ | □なし　□あり | |
| | 味・におい気になるか？ | □ならない　□なる | |
| 服用の煩雑さ | 剤数・回数などに問題は感じているか？ | □なし　□あり | |
| 副作用 | 副作用の経験は？ | □なし　□以前あり　□今もあり | |
| アレルギー | アレルギーは？ | □なし　□あり | |
| 禁忌薬 | 禁忌薬はあるか？ | □なし　□あり | |
| サプリメント | 健康食品・サプリメントの摂取はあるか？ | □なし　□あり | |
| 市販薬 | 市販薬の使用は？ | □なし　□あり | |
| コンプライアンス | 飲み忘れることはあるか？ | □なし　□あり | |
| お薬手帳 | お薬手帳は持参しているか？ | □あり　□あるが持参なし　□なし | |
| 周囲の把握・協力 | 身近に薬について把握している人はいるか？ | □あり　□なし | |
| 薬に対する偏見 | 薬対して不安・思い込みはあるか？ | □なし　□あり | |
| 薬に対する要望 | 薬に対して何か要望はあるか？ | □なし　□あり | |
| その他 | | | |

別紙1 記入例

**評価用紙**　　患者名　　○○ ○○ 様　　回答者　　○○

| 項目 | 質問 | 回答 | コメント |
|---|---|---|---|
| 薬の管理者 | 薬は誰が管理しているか？ | □本人　■家族<br>□その他（　　） | 母 |
| 薬の理解度 | 薬効は？ | □良好　■やや良好　□半分ほど<br>□やや不良　□不良 | |
| | 用法は？ | ■良好　□やや良好　□半分ほど<br>□やや不良　□不良 | |
| 投与法 | 剤形は？ | ■錠剤のみ　□錠剤＋粉薬<br>□粉薬（液剤）のみ　□内服なし | |
| | 投与経路は？ | ■経口　□経鼻<br>□胃瘻　□その他 | |
| | 薬の服用に介助を必要としているか？ | ■不要　□必要 | |
| | 投与法にこだわりはあるか？ | ■なし　□あり | |
| 飲みにくさ | 飲こみにくさはあるか？ | □なし　■あり | 錠剤が大きいので飲みにくい |
| | 味・におい気になるか？ | ■ならない　□なる | |
| 服用の煩雑さ | 剤数・回数などに問題は感じているか？ | ■なし　□あり | |
| 副作用 | 副作用の経験は？ | □なし　■以前あり　□今もあり | 1ヶ月前に |
| アレルギー | アレルギーは？ | ■なし　□あり | |
| 禁忌薬 | 禁忌薬はあるか？ | ■なし　□あり | |
| サプリメント | 健康食品・サプリメントの摂取はあるか？ | □なし　■あり | 青汁 |
| 市販薬 | 市販薬の使用は？ | ■なし　□あり | |
| コンプライアンス | 飲み忘れることはあるか？ | ■なし　□あり | |
| お薬手帳 | お薬手帳は持参しているか？ | ■あり　□あるが持参なし　□なし | |
| 周囲の把握・協力 | 身近に薬について把握している人はいるか？ | ■あり　□なし | ヘルパー |
| 薬に対する偏見 | 薬対して不安・思い込みはあるか？ | ■なし　□あり | |
| 薬に対する要望 | 薬に対して何か要望はあるか？ | □なし　■あり | 大きい錠剤はつぶしてほしい |
| その他 | | | |

別紙2

**薬剤科報告書**

氏名　　様　　年　月　日

1)お薬の内容

| 薬品名 | 用法・用量 |
|---|---|
| | |

2)現状確認

・市販薬　□有 □無 (　　)

・健康食品　□有 □無 (　　)

・薬剤副作用発現歴　□有 □無 (薬品名:　　症状/時期:　　)

・アレルギー歴　□有 □無 (原因物質:　　症状/時期:　　)

・禁忌薬　□有 □無 (薬品名:　　)

・お薬手帳　□有 □無

3)入院時の変更薬品　□有　□無

・追加薬品　□有　□無

| 薬品名: | 用法・用量: |
|---|---|

・用法、用量変更薬品　□有　□無

| 薬品名: | 用法・用量: |
|---|---|

・中止薬品　□有　□無

| 薬品名: |
|---|

4)保管方法

| |
|---|

5)その他 (生活上の注意点等)

| |
|---|
| |

独立行政法人国立病院機構 大牟田病院

薬剤師

別紙2 記入例

**薬剤科報告書**

氏名 ○○ ○○ 様　　○年○月○日

1)お薬の内容

| 薬品名 | 用法・用量 |
|---|---|
| コナン錠 10mg | 朝食後 1 回 1 錠 |
| ワンアルファ錠 0.25μg | 朝食後 1 回 1 錠 |

2)現状確認

・市販薬　□有 ■無 (　　)

・健康食品　■有 □無 ( 青汁　　)

・薬剤副作用発現歴　□有 ■無 (薬品名:　　症状/時期:　　)

・アレルギー歴　□有 ■無 (原因物質:　　症状/時期:　　)

・禁忌薬　□有 ■無 (薬品名:　　)

・お薬手帳　□有 ■無

3)入院時の変更薬品　■有　□無

・追加薬品　■有　□無

| 薬品名: アーチスト錠 2.5mg | 用法・用量: 朝食後 1 回 1 錠 |
|---|---|

・用法、用量変更薬品　□有　□無

| 薬品名: | 用法・用量: |
|---|---|

・中止薬品　□有　□無

| 薬品名: |
|---|

4)保管方法

直射日光、熱、湿気を避けて保管してください。

5)その他 (生活上の注意点等)

今回の入院時にアーチスト錠 2.5mg が追加になっています。飲み始めて、めまい・ふらつき・体がだるいなど普段と異なる症状がある場合は、病院を受診されてください。

錠剤が飲みにくい場合は、錠剤を粉砕したり、他の剤形に変更することもできます。薬剤師に相談されてください。

健康食品の青汁は、お薬によっては飲み合わせが悪い場合があります。新たにお薬が始まるときは、薬局などで確認してください。病院を受診の際は必ずお薬手帳を持参してください。

独立行政法人国立病院機構 大牟田病院

薬剤師 ○○ ○○

# 上記以外のサービス

臨床工学技士、医療ソーシャルワーカー
人工呼吸器の自宅での使用における本人及び家族の技術的習熟度の向上、滞りのない病診連携においてこれらの職種の重要性は言及するまでもないが、本ポートサービスにおいては常に全ての患者において関わるというわけではないことより、本サービスにおいては准メンバーとして協力を仰いでいます。

# さいごに

本マニュアルは、当初本病院のスタッフの経験の差を縮めることができないかということで、作り始めたものでした。これを国立病院機構刀根山病院　名誉院長　神野　進先生に助言をいただき、広く活用できるものを目標に作ってまいりました。

また、今回の改定に当たり、多くの方に助言をいただきましたことを、ここに記させていただきます。

2011年12月

本冊子につきまして、ご不明な点がございました際には、いつでも結構ですので、担当者までご連絡いただければ幸いです。

# 筋ジストロフィー・ポートサービス実施要項
# 第２版

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療育指導室　児童指導員　落合　亮介

13

# Database of Wards for Patients with Muscular Dystrophy in Japan

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*Japan*

## 1. Introduction

Twenty-seven hospitals in Japan 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 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 between 1999 and 2010 in order to evaluate the medical condition of inpatients with muscular dystrophy in Japan.

## 2. Subjects and methods

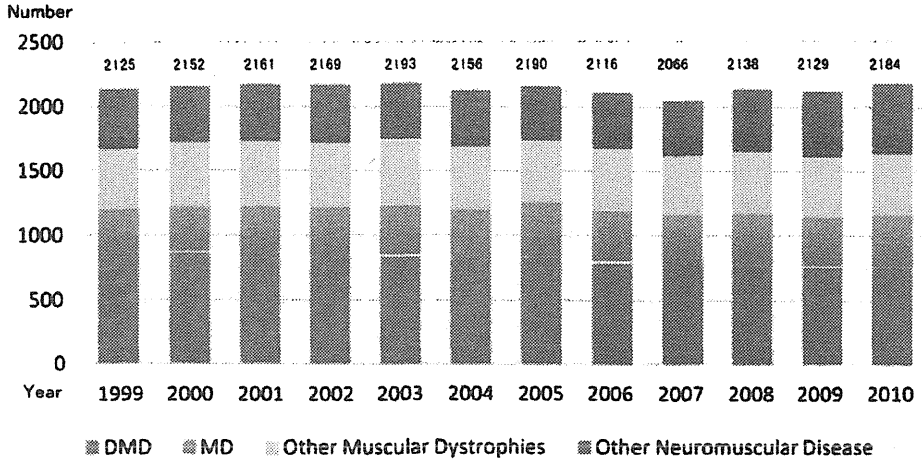
The database includes numbers of inpatients, gender, age, diagnosis, respiratory condition, nutritional state, number of death cases, causes of death, and other relevant findings from data collected annually on October 1 every year since 1999. We examined these data using longitudinal and horizontal analyses.

## 3. Sequential changes in total numbers of inpatients treated at muscular dystrophy wards of National Hospital Organization and National Center of Neurology and Psychiatry

The total numbers of inpatients treated at the muscular dystrophy wards of the National Hospital Organization and National Center of Neurology and Psychiatry were quite consistent during the examination period. The lowest number of inpatients was 2066 in 2007 and the highest was 2193 in 2003 (Fig. 1).

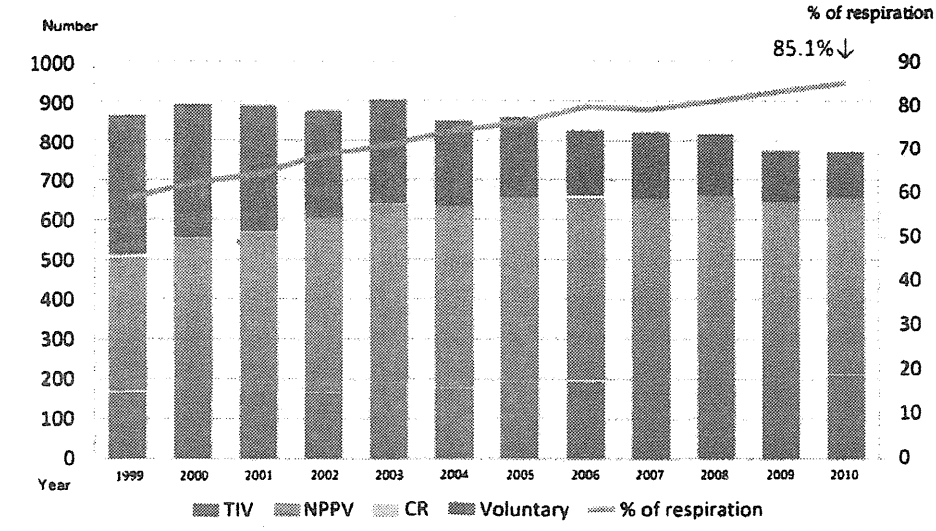
### 3.1 Details regarding number of inpatients

The number of inpatients with Duchenne muscular dystrophy gradually decreased (882~770) every year (Fig. 2), whereas that of those with myotonic dystrophy gradually increased (327~411) (Fig. 3). The numbers of inpatients with other types of muscular dystrophy, such




"Other muscular dystrophies" includes Becker muscular dystrophy, Fukuyama congenital muscular dystrophy, limb-girdle muscular dystrophy, facio-scapulo-humeral muscular dystrophy, Ullrich muscular dystrophy, and others.
"Other neuromuscular disease" includes amyotrophic lateral sclerosis, spinal muscular atrophy, hereditary sensory motor neuropathy, congenital myopathy, and others.
DMD, Duchenne muscular dystrophy; MD, myotonic dystrophy

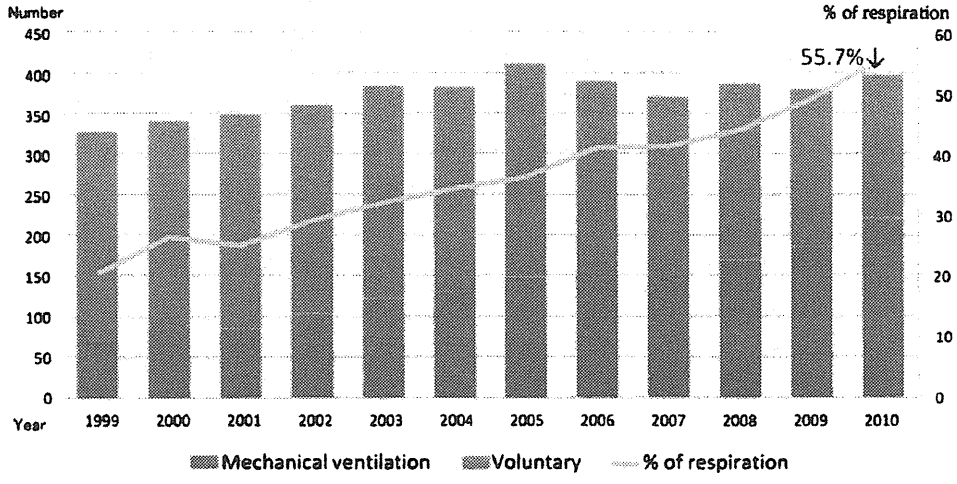
Fig. 1. Total numbers of inpatients in muscular dystrophy wards of National Hospital Organization and National Center of Neurology and Psychiatry.




The number with Duchenne muscular dystrophy has gradually decreased every year.
TIV,tracheostomy intermittent ventilation; NPPV, non-invasive positive pressure ventilation

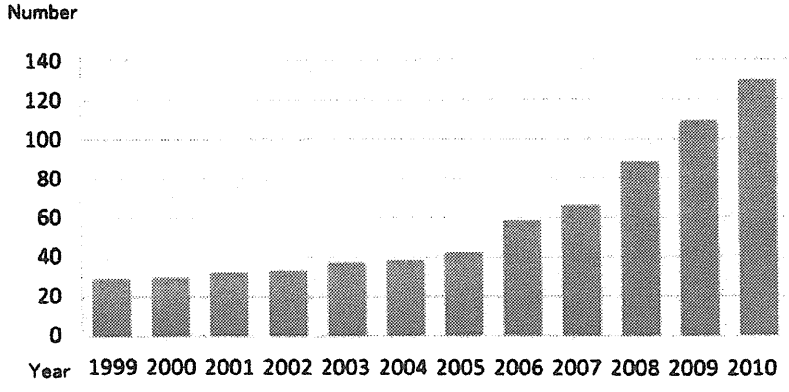
Fig. 2. Sequential changes in number of inpatients with Duchenne muscular dystrophy and rate of mechanical ventilation dependence.

as Becker muscular dystrophy (94~105), Fukuyama congenital muscular dystrophy (50~64), limb-girdle type muscular dystrophy (185~216), and facio-scapulo-humeral muscular dystrophy (64~72) showed some fluctuations. Inpatients with spinal muscular atrophy showed a gradual decreasing tendency from 73 in 1999 to 56 in 2010, while those with amyotrophic lateral sclerosis increased every year from 29 to 132 (Fig. 4). Other diseases encountered in these patients included congenital metabolic disease, mitochondrial disease, various types of myopathy, peripheral nerve disease, bone disease, chromosomal abnormalities, spinocerebellar ataxia, neonatal period disease sequelae, infectious diseases, and others, though their numbers were small and equalled around 10% of all diseases.




The number with myotonic dystrophy has gradually increased every year.

Fig. 3. Sequential changes in number of inpatients with myotonic dystrophy and rate of mechanical ventilation dependence.

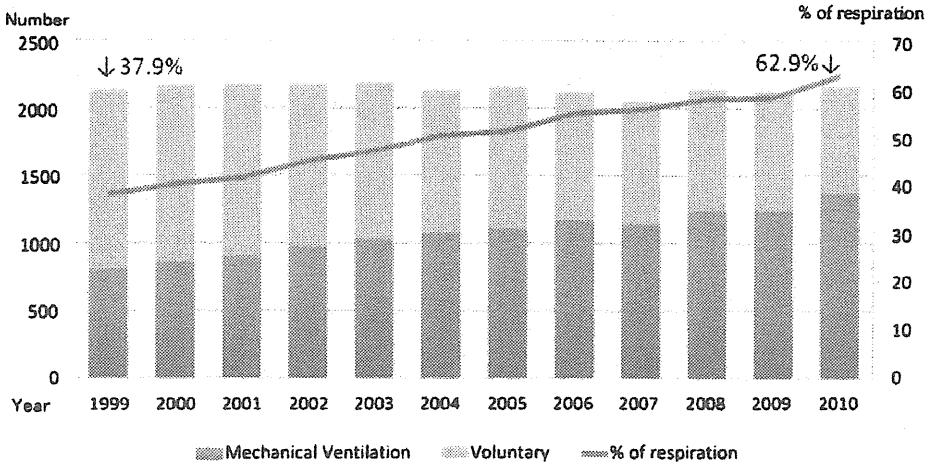



The number with amyotrophic lateral sclerosis has gradually increased every year.

Fig. 4. Sequential changes in number of inpatients with amyotrophic lateral sclerosis.

### 3.2 Sequential changes in respiratory care for inpatients and rate of mechanical ventilation dependence

The rate of mechanical ventilation use in 1999 was 37.9%, which gradually increased to 62.9% in 2010 (Fig. 5), while that for Duchenne muscular dystrophy patients in 1999 was 58.7% and gradually increased to 85.1% in 2010 (Fig. 2). Although the total number of inpatients with Duchenne muscular dystrophy gradually decreased, cases of non-invasive ventilation gradually increased and tracheostomy cases were also slightly increased. The rate of mechanical ventilation use for myotonic dystrophy patients in 1999 was 20.3%, which gradually increased to 55.7% in 2010 (Fig. 3).




The rate of mechanical ventilation use in 1999 was 37.9%, which gradually increased to 62.9% in 2010.

Fig. 5. Sequential changes in respiratory care for inpatients and rate of mechanical ventilation dependence.

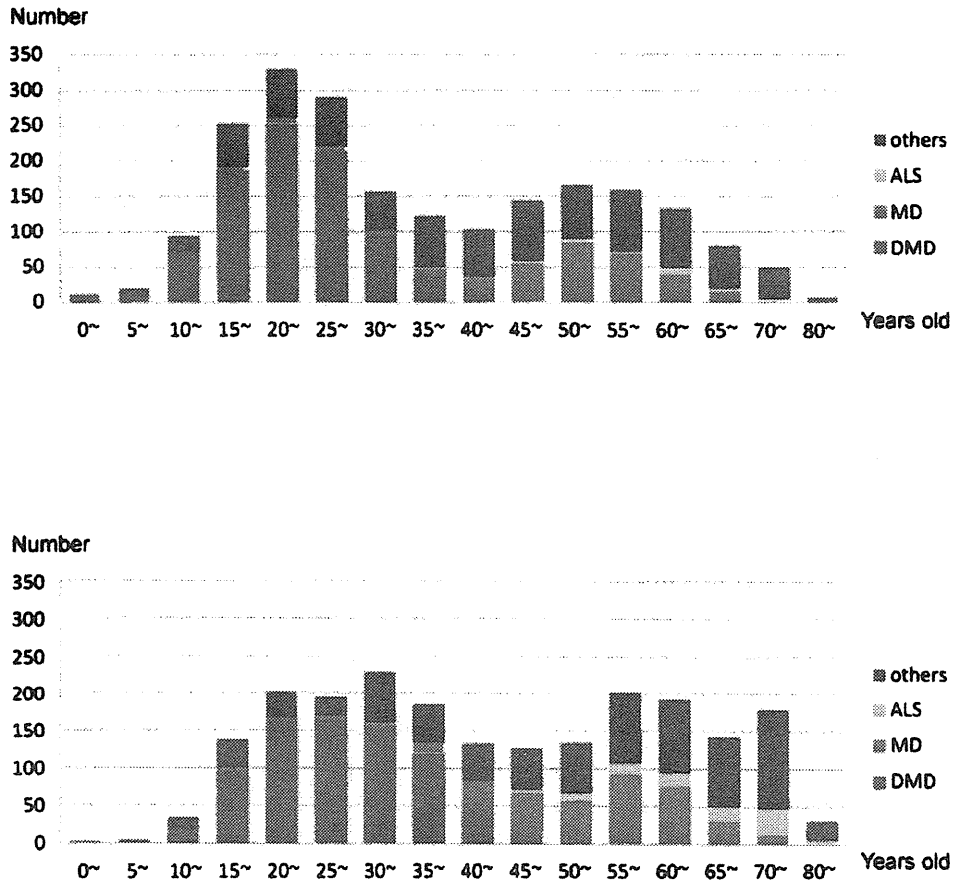
### 3.3 Analysis of mean age of inpatients

#### 3.3.1 Changes in age distribution of inpatients in muscular dystrophy wards

The age distribution of inpatients in muscular dystrophy wards in 1999 showed 2 peaks. Those with Duchenne muscular dystrophy largely constituted the younger age peak in the 20s, while those with myotonic dystrophy larger constituted the older age peak in the 50s. These age peaks shifted to a higher range and became slightly flattened in 2009 (Fig. 6).

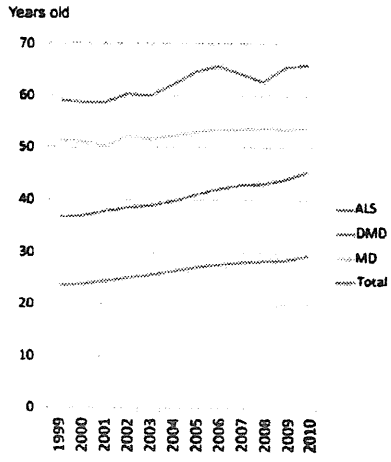
#### 3.3.2 Sequential changes in mean age of inpatients

The mean age of the inpatients in 1999 was 36.6 years old, which gradually increased to 45.3 years old in 2010. That of Duchenne muscular dystrophy patients in 1999 was 23.6 years old, which gradually increased to 29.4 years old in 2010, while that of myotonic dystrophy patients changed only slightly from 51.4 years old in 1999 to 53.6 years old in 2010 (Fig. 7).




Upper: 1999. Lower: 2009. The age distribution of inpatients in muscular dystrophy wards shifted to a higher range over time.

Fig. 6. Changes in age distribution of inpatients in muscular dystrophy wards.




The mean age of the inpatients was gradually increased. DMD, Duchenne muscular dystrophy; MD, myotonic dystrophy; ALS, amyotrophic lateral sclerosis

Fig. 7. Sequential changes in mean age of inpatients.

Gradual changes in age distribution of inpatients with Duchenne muscular dystrophy was observed. The age peak in 1999 shifted to a higher range and became slightly flattened in 2009 (Fig. 8).

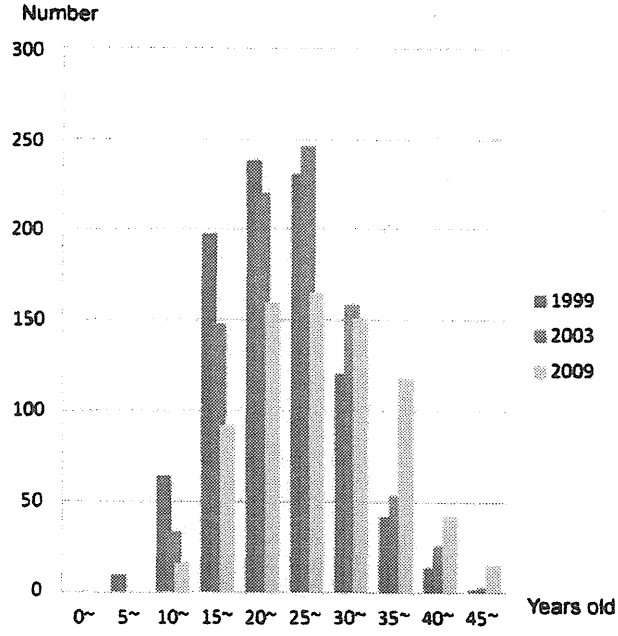
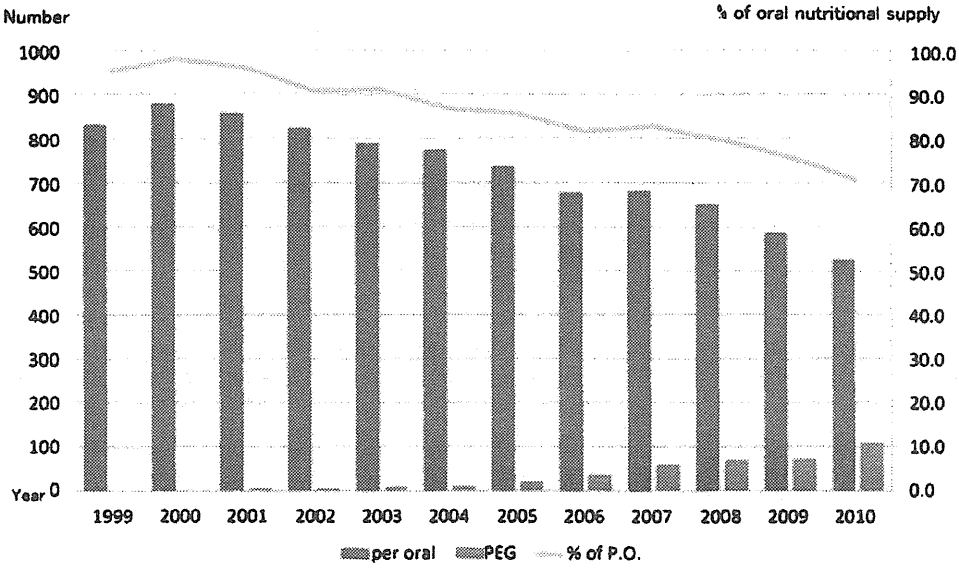



Fig. 8. Changes in age distribution of inpatients with Duchenne muscular dystrophy.

### 3.4 Sequential changes in numbers of patients receiving oral nutrition and those with Duchenne muscular dystrophy who underwent a percutaneous endoscopic gastrostomy

The proportion of patients with Duchenne muscular dystrophy receiving oral nutrition in 1999 was 95.1%, which gradually decreased to 70.6% in 2010. In contrast, the number who required tube feeding, including a nasal nutrition tube and undergoing a percutaneous endoscopic gastrostomy, gradually increased to 107 in 2010.




PEG, percutaneous endoscopic gastrostomy

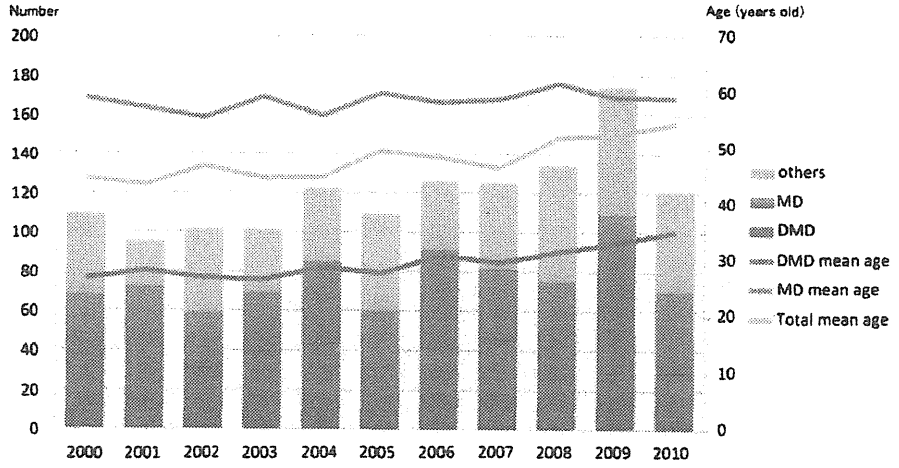
Fig. 9. Sequential changes in numbers of Duchenne muscular dystrophy patients and those who underwent an endoscopic gastrostomy patients receiving oral nutrition.

### 3.5 Death case analysis

The total number of deaths reported from 2000 to 2010 was 1307, which ranged from 95-174 annually in a variable pattern (Fig. 10). The number of Duchenne muscular dystrophy patients who died was 409, while that of myotonic dystrophy patients was 363.

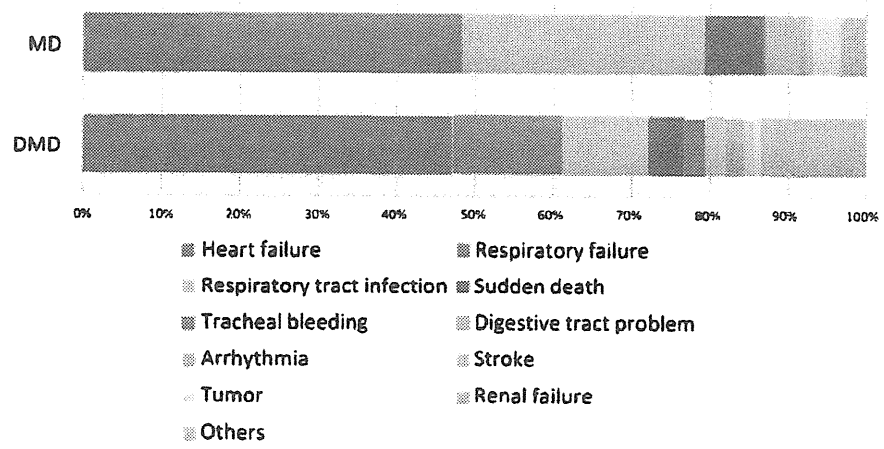
The mean age of death among Duchenne muscular dystrophy patients was 26.7 years old in 2000, which gradually increased to 35.1 years old by 2010. On the other hand, the mean age of death for myotonic dystrophy patients was 59.0 years old in 2000 and 59.1 years old in 2010, which was not significantly different (Fig. 10).

The most frequent cause of death among Duchenne muscular dystrophy patients was heart failure, accounting for 47%. As for myotonic dystrophy patients, the most frequent cause was respiratory disorders, such as respiratory failure and respiratory tract infection, which accounted for 64% (Fig. 11).




DMD, Duchenne muscular dystrophy; MD, myotonic dystrophy

Fig. 10. Sequential numbers of deaths and mean age at death reported to the database.



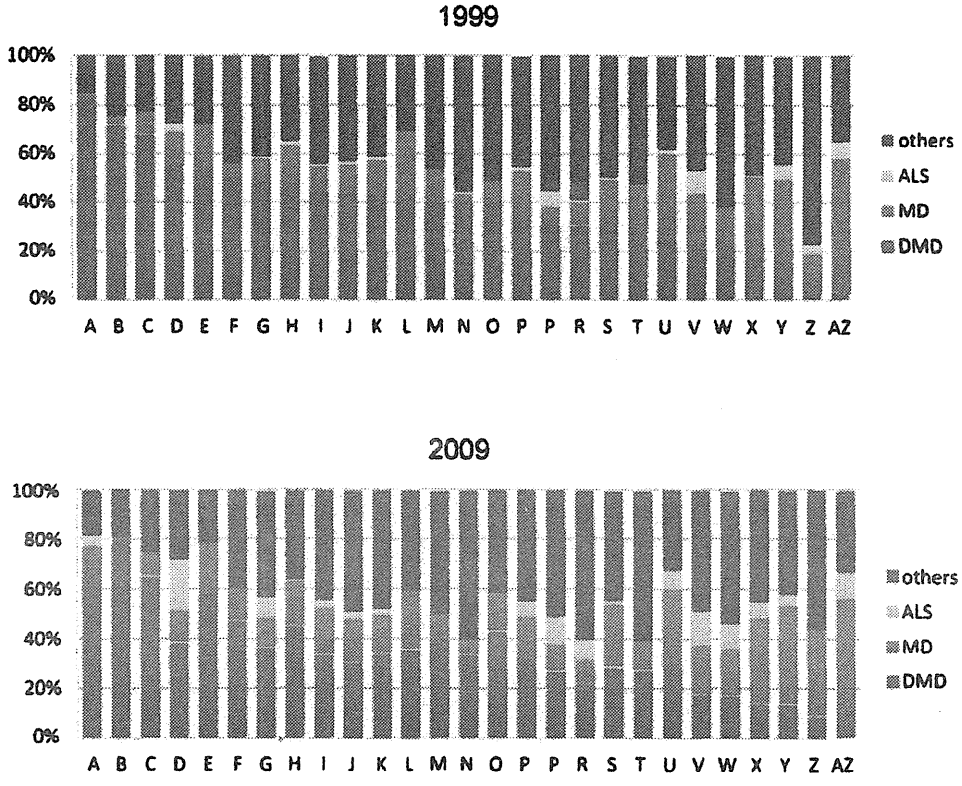

The most frequent cause of death among Duchenne muscular dystrophy patients was heart failure. In contrast, that of myotonic dystrophy patients was respiratory disorder.
DMD, Duchenne muscular dystrophy; MD, myotonic dystrophy

Fig. 11. Causes of death among Duchenne muscular dystrophy and myotonic dystrophy patients (2000~2010).

### 3.6 Proportional changes in numbers of inpatients in muscular dystrophy wards of each institution

Twenty-seven hospitals in Japan specialize in treatment of muscular dystrophy patients are not same in terms of types of muscular dystrophy of inpatient, disease severity, and actual care. Fig. 12 shows the proportion of inpatients by each institution. The upper figure, which

shows the proportion in 1999, is arranged according to rate of Duchenne muscular dystrophy inpatients. There were significant differences in regard to the proportion of inpatients among the institutions in 1999, which changed over time. In 2009, the proportion of inpatients with amyotrophic lateral sclerosis was notable.



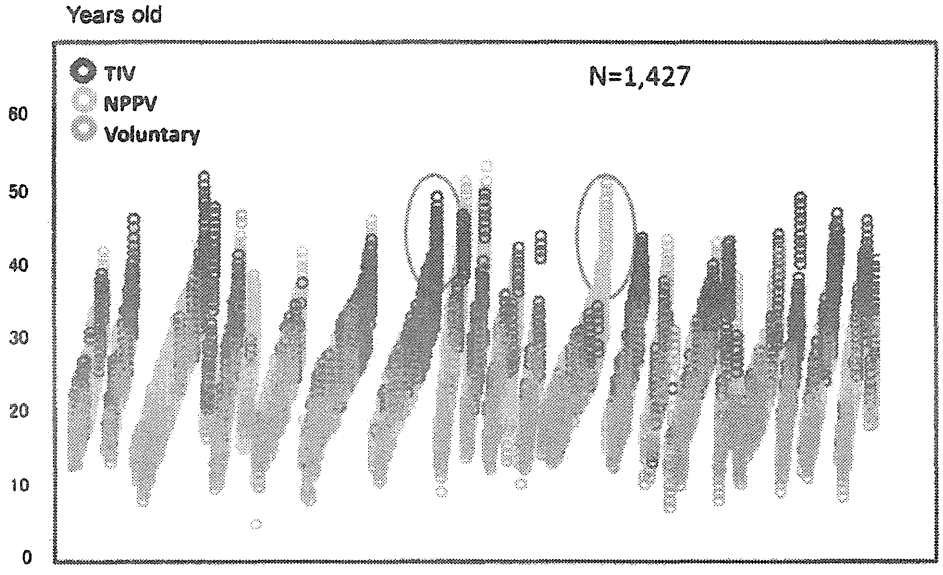

Upper: 1999. Lower: 2009. A~AZ represent the individual institution. Institute AZ, which had lowest rate of DMD patients among institutions in 1999, has no DMD patient in 2009.
DMD, Duchenne muscular dystrophy; MD, myotonic dystrophy; ALS, amyotrophic lateral sclerosis

Fig. 12. Changes in proportions of inpatients in muscular dystrophy wards of each institution

### 3.7 Sequential changes in respiratory conditions of Duchenne muscular dystrophy patients at each institution (1999~2009).

The total number of Duchenne muscular dystrophy patients treated from 1999 to 2009 was 1427. The changes in motor function of the patients were nearly uniform, whereas the therapeutic respiratory conditions varied among the institutions.

Figure 13 presents the respiratory conditions of the patients for the 11-year period from 1999 to 2009. In the 10s, almost patients keep voluntary respiratory function. In the 20s, various respiratory patterns are observed, which seem not to be different among the institutions. In more than 30s, there were apparent differences among the institutions. Some institutes have no tracheostomy older patients, which generation is generally supposed not to be compensated by non-invasive positive pressure ventilation and use tracheotomy ventilation.




Each cluster indicates a single institution. The vertical axis indicates the course of a single Duchenne muscular dystrophy patient. The respiratory conditions of older patients differed among the institutions. For example, the left oval indicates a tracheostomy case and the right oval a non-invasive positive pressure ventilation case.
TIV,tracheostomy intermittent ventilation; NPPV, non-invasive positive pressure ventilation

Fig. 13. Sequential changes in respiratory conditions of Duchenne muscular dystrophy patients treated at each institution (1999~2009).

## 4. Conclusion

Wards for patients with muscular dystrophy were originally established in Japan in 1964 and then gradually expanded throughout the country. As a result, approximate 2500 beds are now provided among 27 institutions. In the early days, many of the patients were boys with Duchenne muscular dystrophy, who received education in schools near the hospital where they received care. However, over time, regular public elementary and junior high schools began to accept disabled children, and such patients were then able to receive an education at schools in their home town. Thus, cases of admission for the purpose of education gradually decreased.

On the other hand, progress in therapeutic strategies for respiratory failure (American Thoracic Society Documents, 2004), heart failure (Ishikawa, 1999; Matsumura, 2010) and other complications associated with muscular dystrophy prolonged the life span of affected individuals (Bushby 2010a, b). Now, most inpatients admitted to a muscular dystrophy ward have a severe general condition and many are assisted by mechanical ventilation (Tatara, 2008). In addition, in terms of nutritional control (American Thoracic Society Documents, 2004; Bushby 2010b), the number of percutaneous endoscopic gastrostomy patients with Duchenne muscular dystrophy has gradually increased.

Thus, the age and disease severity of inpatients have been gradually progressed with this changing environment. And social welfare systems related to muscular dystrophy wards in Japan also have been changing during this research. The social role of wards for inpatients with muscular dystrophy has been changing. The gradual increase of number of inpatients with amyotrophic lateral sclerosis means that the ward for patients with muscular dystrophy is no longer only for patients with muscular dystrophy. Present wards have purpose for care and treatment for severe disabilities, not limited to patients with muscular dystrophy.

There are some reports concerned with prognosis of patients with Duchenne muscular dystrophy from single institution belonging to the National Hospital Organization (Ishikawa, 2011; Matsumura, 2011). Just as these reports, we showed the increasing mean age of death among Duchenne muscular dystrophy patients. Although the most frequent cause of death among Duchenne muscular dystrophy patients was heart failure, the progression for cardioprotection therapy to cardiomyopathy (Ishikawa, 1999; Matsumura, 2010) improved the prognosis.

However, the present findings showed that there are apparent differences in regard to the proportion of inpatients and therapeutic conditions among institutions. Hereafter, these differences will be more remarkable. So far almost same therapy has been offered among the National Hospital Organization and National Center of Neurology and Psychiatry. However, these conditions will not continue and may influence the prognosis of patients with muscular dystrophy in Japan.

Social role of wards for patients with muscular dystrophy at establishment, offering patients with muscular dystrophy opportunities of education and treatment, has changed into offering severe disabilities care and treatment. We should consider how to manage these conditions.

## 5. Acknowledgments

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Institutions specializing in muscular dystrophy treatment in Japan (Fig.14)

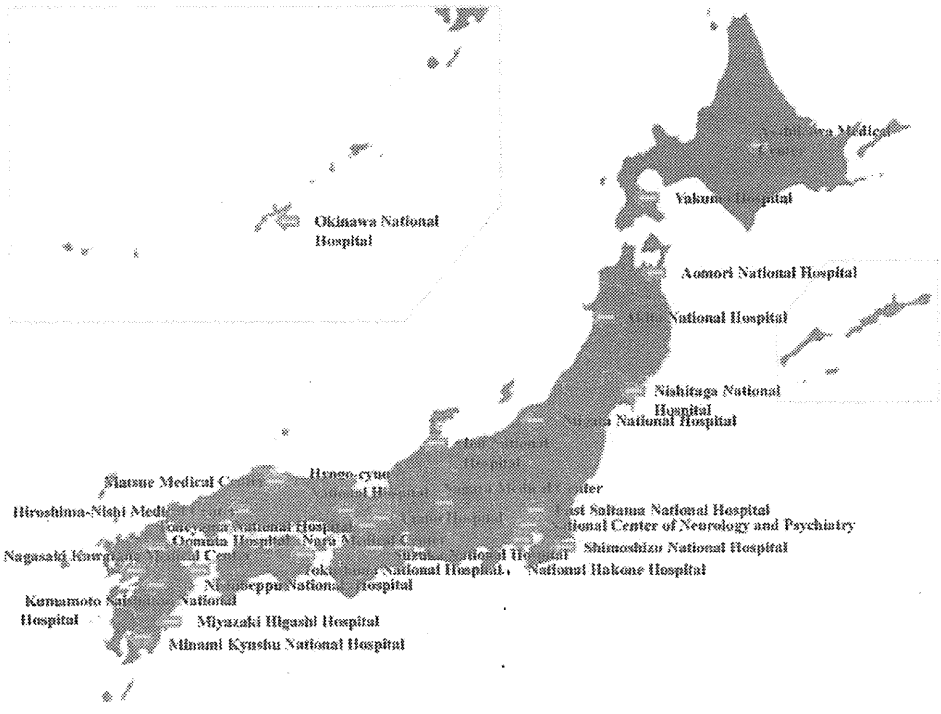


Fig. 14. Institutions specializing in muscular dystrophy treatment in Japan