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Original article

Long-term follow-up of dysphagia in adult patients with Duchenne muscular dystrophy

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ABSTRACT

Background: Dysphagia is a well-known dysfunction in patients with Duchenne muscular dystrophy (DMD), but long-term follow-up of this symptom has not been reported. The aims of this study were to investigate swallowing status, prospectively study the prognosis of swallowing function in adult patients with DMD over the course of 6 years, and to identify clinical factors related to prognosis.

Methods: Participants were 81 patients with DMD. Swallowing status was assessed using the Neuromuscular Disease Swallowing Status Scale (NdSSS) every 2 years for 6 years.

Results: NdSSS was related to age, functional ability assessed using the Swinyard staging and the status of ventilator dependence. Median NdSSS changed from 6 to 4, 4 and 3 over the 4 time points. Number of patients with tube feeding increased by 5–10% every 2 years. Initial NdSSS was a predictor of future swallowing status.

Conclusion: Swallowing status declined over time in patients with adult DMD, with initial NdSSS as a predictor.

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1. Introduction

Duchenne muscular dystrophy (DMD) is a recessive X-linked genetic disorder that affects approximately one in 3500 live male births. In DMD, the patient is unable to produce the protein dystrophin as a result of a large variety of mutations/deletions of the dystrophin gene, found on chromosome Xp21.¹ The condition is characterized by progressive skeletal, respiratory, and cardiac muscle weakness, leading to severe physical

dependence. Loss of ambulation usually occurs between 10 and 11 years old, and death usually occurs in the early 20s, typically from respiratory complications.² However, recent studies have suggested that survival is increasing due to advances in care, specifically in proactive respiratory management.^{3–6}

Historically, swallowing issues in DMD have not gained a great deal of attention. However, as the life expectancy of patients has gradually increased alongside improvements in medical management, concerns about dysphagia warrant immediate attention. Dysphagia is common and might be

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influenced by the severity of impairments in physical function, in addition to age, in patients with DMD. Understanding feeding problems is very important not only for quality of life,^{7,8} but also for nutritional intake and life expectancy related to respiratory complications.^{9,10} However, few reports have examined feeding status, especially for adult patients, and no studies appear to have reported long-term follow-up of dysphagia in DMD. Information about feeding status and changes over time in patients with DMD is necessary to develop therapeutic strategies.

We aimed to investigate swallowing status and to prospectively study the prognosis of swallowing function in adult patients with DMD over 6 years, and to identify clinical factors associated with an increased risk of swallowing dysfunction.

2. Materials and methods

2.1. Participants

The inclusion criterion was a diagnosis of DMD based on clinical history, physical findings, genetic evaluation, or laboratory studies that included muscle enzymes, electromyography, DNA analysis, or muscle biopsy. Participants were 81 consecutive inpatients with DMD who were hospitalized at Higashi-Saitama National Hospital for long-term care or admitted for short-term evaluation in January 2010. No patients declined to participate in the assessments. The study protocol was approved by the institutional ethics review board (#17-3) and was registered to the University Hospital Medical Information Network (UMIN) clinical trial registry (UMIN 000029939). The study was performed in accordance with the Declaration of Helsinki.

2.2. Study 1: cross-sectional study of dysphagia in patients with DMD

The first study was conducted in January 2010 and employed a cross-sectional design to investigate swallowing status in adult DMD patients. The aims of the study were to examine relation-ships between swallowing status in daily life with age, physical function ability, nutritional status, cardiac function and respiratory function. All 81 patients received the following evaluations.

2.3. Study 2: time course of swallowing status in patients with DMD

The second study employed a longitudinal prospective design to investigate the course of dysphagia in patients with DMD over the course of 6 years, from January 2010 (baseline) to January 2016. The aims of the study were to examine changes in swallowing status and the proportion of tube feeding in adult patients with DMD. We investigated factors predictive of tube feeding. For patients who died within the 6-year study period, we investigated causes of death.

2.4. Assessments

2.4.1. Swallowing status

Swallowing status in daily life was evaluated using the Neuromuscular Disease Swallowing Status Scale (NdSSS).¹¹ The NdSSS is an eight-stage scale that was developed to evaluate dysphagia in patients with progressive neuromuscular diseases, offers acceptable reliability, validity, and responsiveness,¹¹ and has been used for DMD patients. All patients were evaluated swallowing performance using videofluorography and nutrition status was evaluated with blood tests and transition of body weight more than once a year. The level of feeding and use of supplemental nutritions are determined based on the conference of physicians, physiatrists, nurses, speech therapists and nutritionists.

2.4.2. Physical functional ability

Physical functional ability was evaluated using Swinyard's Criteria for Rating Eight Stages of Functional Ability.¹² Swinyard's criteria are based on the pattern and method of ambulation, as well as the ability to perform activities of daily living. Patients were classified into 8 stages, from stage 1 (mildest: patient can ambulate and perform evaluation activities adequately) to 8 (most severe: bedridden status).

The NdSSS and Swinyard's Criteria for Rating Eight Stages of Functional Ability were assessed by the well trained physiatrist, who had engaged in the treatment of patients with neuromuscular diseases for more than 10 years.

2.4.3. Nutritional status

Body mass index (BMI) was calculated as the weight in kilograms divided by the square of the height in meters. Height was calculated as the sum of the four parts separated by the following five landmarks: top of the head, acromion process of the shoulder, greater trochanter of the hip, lateral joint line of the knee joint, and bottom of the heel.¹³ Serum albumin level was also obtained from blood tests as a parameter of nutritional status.

2.4.4. Respiratory and cardiac functions

Partial pressures of carbon dioxide and oxygen in arterial blood ($PaCO_2$ and PaO_2 , respectively) and BNP hormone level were obtained from blood tests.

Status of ventilator dependence was scored on a 4-point scale from 0 to 3: 0 (ventilator-independent), 1 (partially noninvasive positive pressure ventilator (NPPV)-dependent), 2 (NPPV-dependent all day), or 3 (tracheostomy positivepressure ventilator (TPPV)-dependent).

2.5. Statistical analyses

2.5.1. Study 1

Correlations between NdSSS and other parameters (age, Swinyard stage, BMI, serum albumin, $PaCO_2$, PaO_2 and status of ventilator dependence) were quantified using Spearman's rank correlation test. After assessing multicollinearity, multiple regression analysis for NdSSS was conducted. Variables related to NdSSS at p < 0.10 in the above analyses were entered.

2.5.2. Study 2

After univariate logistic regression analyses, logistic analyses (forward selection; likelihood ratio, removing terms with P > 0.10) were performed to examine which variables in 2010 were related to NdSSS in 2016. How closely the prediction reflected observed events, was assessed by the Hosmer–Lemeshow test and a calibration plot.

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