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Cardiac and Pulmonary Management Status of Duchenne Muscular Dystrophy in South Korea Based on Data From the National Health Insurance Database

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Background and Purpose The purpose of this study was to determine the cardiac and pulmonary management status of patients with Duchenne muscular dystrophy (DMD) in South Korea based on the Korean National Health Insurance database.

Methods This study used data of patients with code G71.0 in the National Health Information database, and also those with the special case of code V012. Cardiac function was assessed based on whether echocardiography and 24-h electrocardiography were performed, as well as the frequency of these investigations. Furthermore, information on the use of angiotensin-converting enzyme inhibitors (ACEi), angiotensin II receptor blockers (ARB), and beta blockers (BB) in the drug benefit list was checked. Medical charge records were also checked regarding the prescription of respiratory functional assessments and treatments.

Results The diagnostic criteria were met by 479 patients, with 41% of these patients receiving a cardiac evaluation, 29.8% being prescribed ACEi at 14.4±3.7 years of age, 59.5% undergoing pulmonary function tests, and 42.1% received pulmonary rehabilitation. The age at receiving ventilator support was 19.4±2.7 years. The frequency of cardiac and respiratory function tests increased with age, but the interval between tests was longer than the recent DMD care recommendations. The trend of taking ACEi, ARB, and BB for cardiac management in South Korea did not change during the study analysis period.

Conclusions The findings of this study will contribute to recognizing the current status and the importance of applying an anticipatory approach to cardiopulmonary function in DMD patients.

Keywords Duchenne muscular dystrophy; pulmonary function; cardiac function; South Korea.

INTRODUCTION

Life expectancy has been prolonged by adopting a proactive approach to cardiopulmonary management in patients with Duchenne muscular dystrophy (DMD).[1,2] A few decades ago, most patients with DMD died in their 20s from respiratory failure or cardiomyopathy.[3,4] The consensus of starting glucocorticoid treatment in young children with DMD has been shown to preserve physical and respiratory functions.[5,6] In addition, anticipatory therapeutic strategies including part-time noninvasive ventilation use and cardioprotective medications have been applied,[7] and many clinicians manage patients based on the DMD care recommendations that were updated in 2010[8,9] and 2018.[10,11]

According to these guidelines, the early diagnosis and treatment of cardiopulmonary complications is essential to maximizing both survival duration and the quality of life. Echocar-

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diography is recommended until at least an age of 6–7 years, and initiation of angiotensin-converting enzyme inhibitors (ACEi) in asymptomatic males younger than 10 years could improve long-term cardiac outcomes. Annual echocardiography is recommended during the asymptomatic period to promote the early diagnosis of left ventricular dysfunction. From the onset of heart failure symptoms or abnormalities in cardiac imaging, the frequency of assessments should increase at the discretion of the cardiologist.[11] The recent criteria recommend greater anticipatory use of assisted coughing and ventilation in these patients. Several studies investigating the current clinical practice of DMD have used a DMD registry.[12-14] According to the clinical outcomes in DMD from the TREAT-NMD DMD global database,[14] 10.3% of all DMD patients were using assisted ventilation and 50.4% of these patients were aged 20 years and older. Furthermore, 66.5% of patients diagnosed with cardiomyopathy were evaluated for a reduced ejection fraction using periodic echocardiography.

A DMD registry does not exist in most Asian countries. Therefore, a database needs to be established for clinical information on patients with DMD in South Korea. Furthermore, no previous study has examined the frequency of guideline-recommended periodic pulmonary, cardiac, and bone evaluations and treatment. The associated uncertainties increase the risks of both undertreatment and overtreatment, which could confound the clinical prognosis of DMD patients.

Due to the above-described situation, the present study aimed to determine the current pulmonary and cardiac management status of patients with DMD in South Korea based on the Korean National Health Insurance database, as well as to provide fundamental data for creating and standardizing the national guidelines for DMD treatment for use by clinicians in South Korea.

METHODS

Identification of subjects

This study extracted claims data from the National Health Information database on insurance benefits provided between 2002 and 2018. The study used data of patients with code G71.0 in that database, as well as those with the special case of code V012, according to the tenth revision of the International Classification of Diseases. To screen patients with DMD from the diagnosis data, the operational definition was set as a male patient younger than 40 years who had been diagnosed before the age of 10 years. To exclude other types of muscular dystrophy (MD), patients who had been prescribed steroids were selectively extracted according to the recent recommendation on the current status of steroid prescriptions for DMD patients in South Korea[10] to construct a database. In addition, patients who received ventilator support at younger than 10 years and were prescribed steroids or cardiac medications prior to the diagnosis were excluded from this study, since they were determined as having a weak association with the typical clinical course of DMD (Fig. 1).

Data acquisition and analysis

To ensure the validity of the extracted data, patients were extracted from the database using the codes for specific local tertiary hospitals and nursing facilities established under special purposes that were considered research institutions in 2018. The number of these patients was compared with the actual number of patients who were screened with the diagnostic code for DMD and who received steroid treatment.

Cardiac function was assessed based on whether echocardiography and 24-h electrocardiography were performed as well as the frequency of these investigations. Furthermore, information on the use of ACEi, angiotensin II receptor blockers (ARB), and beta blockers (BB) in the drug benefit list provided by the National Health Insurance database was checked.

The medical charge record was checked for prescriptions of respiratory functional assessments and treatments, and their frequency, in addition to checking the benefits payment table regarding receiving ventilator support and the onset of its use.

The test and prescription codes for cardiopulmonary management analyzed in this study are presented in Supplemen-

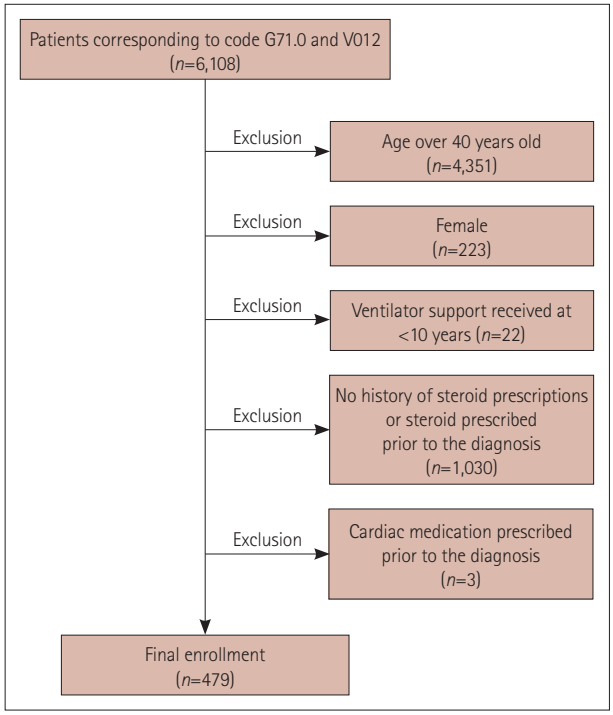

Fig. 1. Flow diagram of screening the number of patients with Duchenne muscular dystrophy using corticosteroids.

tary Table 1 (in the online-only Data Supplement).

Ethics statement

The study was approved by the Institutional Review Board (IRB) of Pusan National University Hospital (IRB No.1907-008-080) and the National Health Insurance Service of Korea (REQ0000030402). The requirement for informed consent was waived because secondary data were used.

RESULTS

From 2002 to 2018, 479 patients met the diagnostic criteria for inclusion in this study. For validation, the data of 25 patients in 2018 were extracted from specific local tertiary hospital facilities (the investigators' institutions). Reviews of medical records identified 26 DMD patients who received steroid treatment at our hospital in 2018.

Demographics characteristics including for mortality and the causes of deaths are summarized in Supplementary Figs. 1 and 2 (in the online-only Data Supplement). Among our patients, the age at which wheelchair support was received between 2016 and 2018—corresponding to the age at loss of ambulation—was 11.9±3.4 years (mean±standard deviation). Table 1 lists the proportions of cardiac and respiratory evaluations and management, where 41% of the patients received cardiac evaluation and 29.8% were prescribed ACEi at the age of 14.4±3.7 years. In total, 25.6% and 9.3% of patients had been prescribed BB and ARB at 13.2±9.5 and 16.4±4.2 years of age, respectively. There was no trend in the change in the initial prescription of cardiac medication over time (Fig. 2). Furthermore, 59.5% of patients underwent pulmonary function tests and 42.1% received pulmonary rehabilitation. The age at receiving ventilator support was 19.4±2.7 years. The intervals of cardiac and respiratory function evaluations according to age are shown in Fig. 3. The time interval of each cardiac and respiratory function test decreased with age and over time (Table 2). The proportion of patients aged <10 years receiving ACEi increased annually, and the trend in this proportion over time is presented in Table 3.

Table 1. Cardiac and respiratory evaluation and management in patients with Duchenne muscular dystrophy

Parameter	Value (*n*=479)
Cardiac evaluation and management	
Echocardiography	197 (41.1)
24-h electrocardiography	199 (41.5)
ACEi prescription	143 (29.8)
Age at ACEi prescription (yr)	14.4±3.7
BB prescription	123 (25.6)
Age at BB prescription (yr)	13.2±9.5
ARB prescription	45 (9.3)
Age at ARB prescription (yr)	16.4±4.2
Respiratory evaluation and management	
Pulmonary function test	285 (59.5)
Pulmonary rehabilitation	202 (42.1)
Ventilator support	82 (17.1)
Age at receiving ventilator support (yr)	19.4±2.7

Data are *n* (%) or mean±SD values. Ventilator support data were extracted from the benefits payment table for 2016–2018.
ACEi, angiotensin-converting enzyme inhibitors; ARB, angiotensin II receptor blockers; BB, beta blockers.

DISCUSSION

Respiratory and cardiac complications are major causes of morbidity and mortality in patients with DMD.[15] This is the first database study to investigate the current status of clinical cardiopulmonary management practice in patients with DMD in South Korea. In an expert study of the clinical practice of steroid treatment for DMD patients in 2020, most Korean clinicians responded that steroid treatment was initiated after a disease diagnosis.[16] In the present study, to identify DMD patients among MD patients corresponding to code G71.0 as well as the special case of code V012, only male patients younger than 40 years who were prescribed steroids were included. Since it is possible that applying the above-mentioned criteria resulted in the exclusion of some DMD patients who never received steroid treatment, an epidemiological analysis of DMD was not considered appropriate, and the scope of this study was restricted to determining the clinical practices applied to DMD patients who received steroid treatment. The number of patients extracted from specific medical institutions for validation was similar to the actual number of patients from the applicable hospitals, and analysis was performed with the appropriate groups. In addition,

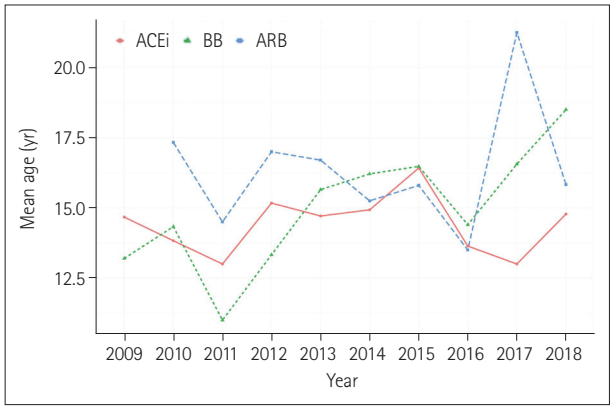

Fig. 2. Age at initial cardiac medication prescription over time. ACEi, angiotensin-converting enzyme inhibitors; ARB, angiotensin II receptor blockers; BB, beta blockers.

exploring the time of receiving wheelchair support in the 3-year benefits payment table revealed that most patients lost their ambulation function after the age of 15 years. Thus, the results of this study were considered by classifying them as nonambulant patients.

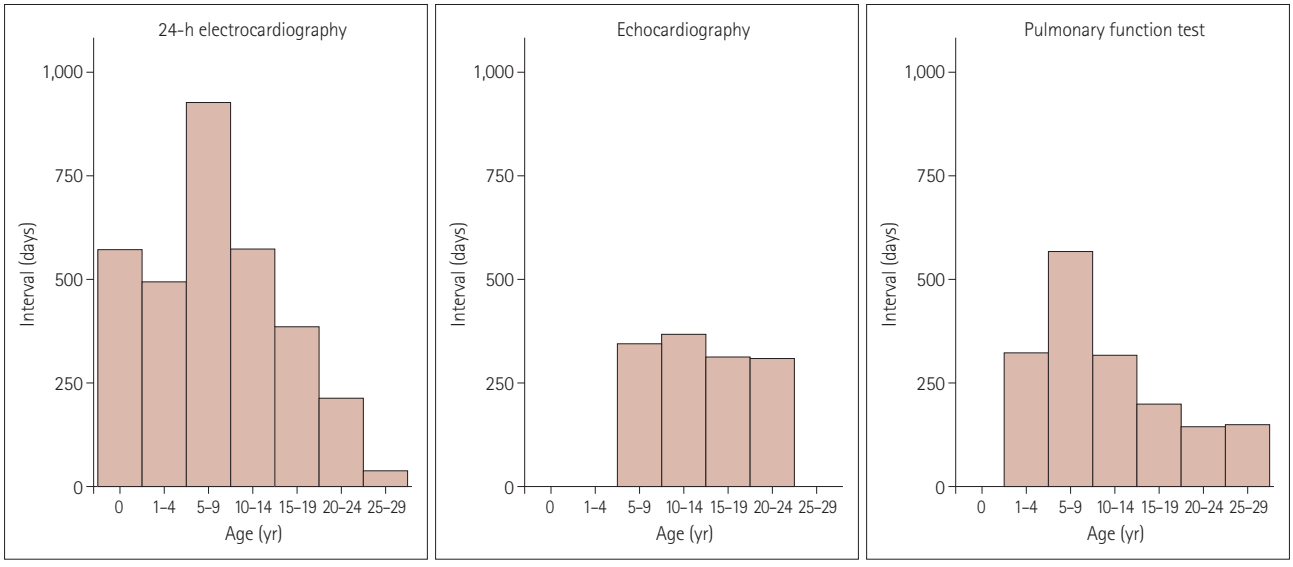

Fig. 3. Intervals of cardiac and respiratory function evaluations.

Table 2. Interval of cardiac and respiratory function evaluations according to age group and year

Age (yr)	2013	2014	2015	2016	2017	2018
Echocardiography						
5–9	511.5±365.5	365.2±106.3	399.3±69.7	384.3±79.7	322.1±64.2	300.3±34.3
10–14	360.8±173.4	390.7±138.3	345.4±84.6	185.5±20.5	193.1±11.7	192.2±29.3
15–19	331.7±94.1	361.3±184.7	270.8±104.6	108.6±79.0	153.6±34.3	142.2±24.2
≥20	380.3±99.4	378.7±97.3	289.4±125.1	162.5±34.6	139.2±45.6	132.5±34.3
24-h electrocardiography						
5–9	157.0±4.24	245.0±106.3	456.6±323.8	495.0±131.5	395.0±201.3	344.0±91.6
10–14	525.6±533.2	381.6±365.5	231.5±317.7	456.6±323.8	251.0±101.5	214.0±80.1
15–19	633.8±689.2	407.6±528.2	407.6±528.2	250.2±246.2	149.2±201.3	86.0±73.9
≥20	1158.5±194.4	438.2±638.9	248.5±362.5	98.7±169.9	89.7±129.1	55.38±44.0
Pulmonary function test						
5–9	220.6±65.6	342.7±390.1	631.8±515.5	467.5±14.8	177.5±201.4	194.2±59.4
10–14	233.2±206.6	243.1±223.8	273.9±252.8	219.6±183.4	217.0±109.7	145.3±60.3
15–19	231.8±282.0	168.5±166.9	204.4±206.8	172.4±117.6	166.9±85.6	109.8±60.6
≥20	153.5±143.9	156.4±213.0	289.4±125.1	156.1±134.1	146.0±96.7	130.9±54.5

Data are mean±SD values for the interval in days.

Table 3. Proportion of ACEi prescriptions according to age group and year

Age (yr)	2009 (*n*=20)	2010 (*n*=24)	2011 (*n*=23)	2012 (*n*=23)	2013 (*n*=36)	2014 (*n*=63)	2015 (*n*=87)	2016 (*n*=92)	2017 (*n*=97)	2018 (*n*=100)
≤4	0 (0)	0 (0)	0 (0)	0 (0)	0 (0)	0 (0)	1 (1.2)	0 (0)	1 (1.0)	1 (1.0)
5–9	3 (15.0)	1 (4.2)	0 (0)	0 (0)	1 (2.8)	2 (3.2)	2 (2.3)	5 (5.4)	6 (6.2)	10 (10.0)
10–14	9 (45.0)	12 (50.0)	7 (30.4)	7 (30.4)	11 (30.6)	21 (33.3)	13 (14.9)	8 (8.7)	7 (7.2)	5 (5.0)
15–19	8 (40.0)	11 (45.8)	16 (69.6)	16 (69.6)	22 (61.1)	30 (47.6)	56 (64.4)	46 (50.0)	46 (47.5)	43 (43.0)
20–24	0 (0)	0 (0)	0 (0)	0 (0)	2 (5.5)	10 (15.9)	15 (17.2)	33 (35.9)	36 (37.1)	38 (38.0)
≥25	0 (0)	0 (0)	0 (0)	0 (0)	0 (0)	0 (0)	0 (0)	0 (0)	1 (1.0)	3 (3.0)

Data are *n* (%) values.
ACEi, angiotensin-converting enzyme inhibitors.

Cardiac management

In terms of anticipatory pulmonary management for patients with DMD, cardiovascular complications are a leading cause of mortality.[17] In particular, there are few cases of excessive physical activity after losing ambulation function, and so symptoms related to heart failure rarely occur; therefore, these complications may be overlooked. Cardiovascular magnetic resonance imaging (MRI) is the diagnostic modality of choice and has recently become covered by National Health Insurance in South Korea. However, sedation is often required due to poor cooperation from pediatric patients, and positioning for MRI is difficult because of the presence of multiple joint contractures in many older patients. Therefore, echocardiography is usually recommended initially for these patients. In general, it is recommended that cardiac imaging studies be performed annually from the age of 6−7 years in asymptomatic cases, since abnormalities may be detected on echocardiography and noninvasive imaging. The frequency of examinations should increase after the onset of symptoms indicative of heart failure or abnormal findings on cardiac imaging, including left ventricular dysfunction.[18] In the present study, 41.1% and 41.5% of patients received echocardiography and 24-h electrocardiography, respectively. Exploring the trend of echocardiography over a 3-year period revealed that the interval tended to shorten with increasing age, which is consistent with the recent age-based recommendations. In 2018, the time interval for echocardiography investigations was 1 year for individuals 10 years or younger and 6-months or shorter for patients older than 10 years, which is consistent with the recent recommendations.[11]

With regard to cardiac management, the frequencies of ACEi, ARB, and BB recommendations were investigated. There was no trend for the changes in age at the initial prescription of cardiac medication over time in our study. In particular, there have been mixed reports on the prophylactic administration of ACEi in children aged <10 years who have no symptoms or abnormalities on cardiac MRI or echocardiography.[19] According to a recent report based on multicenter DMD registry data, the prophylactic administration of ACEi improved overall survival and decreased hospitalization rates due to heart failure.[20] Based on the data collected in the our study, 10% of patients received ACEi at an age of <10-years in 2018, and a trend of an annual increase in this percentage was confirmed for prescriptions of ACEi.

Cardiac rhythm abnormality can develop during the late nonambulatory stage, although the time of initiation and frequency of monitoring have not been established. It is reasonable to initiate annual screening using Holter monitoring after the onset of abnormal left ventricular function or the development of myocardial fibrosis.[21] In the present study, 24-h electrocardiography did not show a decreasing trend in the prescription frequency according to age, but exploring the trend of the most-recent 3 years revealed that the frequency of prescription increased with age, with the interval becoming shorter than 6-months for late nonambulant patients (>15 years old). According to the recommendations, the optimum frequency of 24-h electrocardiography is yet to be established, and is determined by the cardiologist depending on the clinical course of individual patients.[11] In South Korea, it was identified that the test interval for the risk of rhythm abnormalities, which increases with age, has tended to decrease over time.

Respiratory management

Respiratory complications are a major cause of morbidity and mortality in patients with DMD.[22] The care considerations in the revised DMD guidelines of 2018 emphasized the anticipatory management of respiratory function, with the initiation of spirometry recommended at 5−6 years of age, and with this being performed annually during the ambulatory stage and every 6 months in the nonambulatory stage. Assisted coughing and nocturnal ventilation were also reported to decrease respiratory complications and improve the quality of life when initiated from a lower age as an anticipatory intervention.[11] The present study was unable to confirm the frequency of specific pulmonary function tests and rehabilitation criteria, while approximate measurements of pulmonary function were made in almost 60% of the patients. In 2018, the time interval between pulmonary tests was approximately 6 months for patients younger than 10 years, and this frequency increased over time. Only 40% of patients received rehabilitation treatment, which is inconsistent with current recommendations, emphasizing the importance of an anticipatory approach to management including the timely use of ventilation and assisted coughing.[23-25] When we compared the recent changes in the age group that had received ventilator support, the percentage of prescriptions and average age from the 3-year record in this study were insufficient for validating whether it support reflected these recommendations. Since these respiratory prescriptions could decrease respiratory complications and prolong survival,[1,5,26] more anticipatory use of interventions should be endorsed in future DMD care recommendations.

This study has revealed the current status of clinical practice related to the cardiopulmonary management of DMD, and the best of our knowledge, it is the first study of its kind involving the South Korean population. DMD is characterized by a severe progressive course, but this can be altered by appropriate steroid therapy along with respiratory and cardiac management. Although there are recommendations related to DMD management,[11] treatments vary between both

countries and clinicians, which makes it necessary to understand the current status of DMD management in South Korea.

Notwithstanding the above-mentioned strengths, this study also had some limitations. First, not all of the DMD data could be extracted using code G71.0 and the special case of code V012 alone, which were the disease codes applied to the National Health Information database. Therefore, an epidemiological analysis could not be performed using the data collected in this study. However, DMD patients are the only subgroup of MD patients who are prescribed steroids, and considering the recent guidelines for early prescriptions and maintenance doses, it was presumed that relatively few patients would have been excluded. In addition, we only extracted and analyzed cases designated as 'DMD that was prescribed steroid,' which is considered to be more reliable. Nevertheless, the analyzed data would still have excluded patients who had not been prescribed steroids during the data extraction process, and patients with MD (including Becker's MD) were not completely excluded. In recent years, studies (including clinical trials) have investigated the development of therapeutic agents that can change the clinical course of DMD. When a treatment involves a newly developed medication, it is important to maintain physical function as much as possible. Moreover, several studies have included the steroid therapy status of the patients as one of the inclusion criteria, and so it is important to understand the current status of DMD management with steroid therapy in South Korea.

The second limitation is related to receiving ventilator support. This information could only be obtained from 2016 onwards, when payment based on the benefits payment table became possible. Further studies analyzing longer term data would be useful for improving the understanding of the current status of DMD management.

This study has revealed the current status of clinical practice for patients with DMD in South Korea. In addition to adequately reflecting the changing clinical practices in South Korea, it is expected that the findings of this study will contribute to raising awareness about the necessity of establishing a domestic registry in the country for patients with DMD, as well as developing a consensus among clinicians over the long term.

Supplementary Materials

The online-only Data Supplement is available with this article at https://doi.org/10.3988/jcn.2022.18.5.522.

Availability of Data and Material

The datasets generated or analyzed during the study are not publicly available due to the Personal Information Protection Act of Korea and regulations of the National Health Insurance Sharing Service but are available from the corresponding author on reasonable request.

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Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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None

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