

Status of Duchenne Muscular Dystrophy who use ventilator in Korea in 2022

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Introduction

Duchenne muscular dystrophy (DMD) is the most prevalent form of muscular dystrophy, characterized by severe and progressive muscle wasting that results in difficulties with movement and ultimately, the need for assisted ventilation and premature death. With the advent of cardiopulmonary treatments and government support, an increasing number of DMD patients in different age groups are surviving in Korea. DMD patients with reduced muscle strength to the extent of requiring ventilatory support need comprehensive medical management in various fields. We were interested in clinical features and comprehensive medical management of DMD patients requiring ventilatory support. Therefore, we conducted a study to collect detailed clinical data of DMD patients on ventilatory support in Korea and analyzed the findings.

Methods

This is a cross-sectional observational study conducted at the Pulmonary Rehabilitation Center of Gangnam Severance Hospital from January 2022 to December 2022. Duchenne muscular dystrophy (DMD) was confirmed based on genetic findings confirming the presence of dystrophin gene mutation and loss of independent ambulation by the age of 13. Patient conditions and medical treatments, including respiratory status, treatment of cardiomyopathy, nutritional evaluation, presence of central nervous symptoms, and history of spinal surgery, were investigated.

Results

A total of 191 patients who use ventilator were included. The mean age was 29.6 ± 6.2 years, with 4 patients under the age of 20, 108 in their 20s, 65 in their 30s, and 14 over the age of 40 (Fig 1). The average age at initial ventilatory support was 19.5 years, and they had been using ventilator for 10.1 ± 5.1 years. Among them, 178 patients used non-invasive ventilation, while 13 were receiving ventilatory support via tracheostomy. The patients used ventilators for an average of 15.9 hours per day.

A total of 158 patients (82.8%) were receiving medication for cardiomyopathy, and average ejection fraction was $44.8 \pm 14.4\%$. The most commonly used medications were ACE inhibitors or ARBs, and 97 patients were using two or more medications in combination. Two patients were using implantable cardioverter-defibrillators.

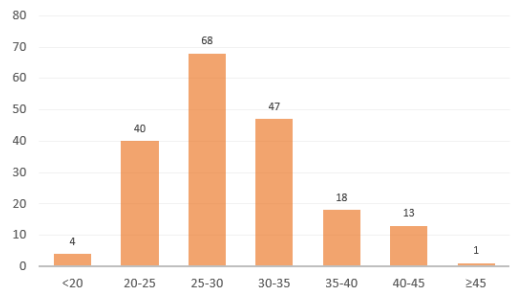
Among the 191 patients, 162 were eating through their mouths only, while 21 were using tube feeding, and 8 were using both methods. Among the patients who were eating through their mouths only, 47 were consuming liquid oral supplements.

A total of 80 patients underwent scoliosis correction surgery. Seven patients (3.7%) exhibited central nervous system symptoms, with 5 experiencing seizures and 2 having intellectual disabilities.

Table 1. Management status of included patients.

Assessment	Characteristics	Mean \pm SD (min-max) or Number (%)
Baseline data	Current Age	29.6 \pm 6.2 (16.2-46.9)
	Age at loss of independent ambulation	10.6 \pm 2.1 (2.0-18.0)
	Height	159.3 \pm 7.9 (130.0-180.0)
	Weight	42.0 \pm 18.4 (19.4-90.0)
	Body mass index	16.5 \pm 5.5 (8.0-37.5)
Respiratory status	Age at initial ventilatory use	19.5 \pm 4.2 (10.7-32.9)
	Tracheostomy	13 (6.8%)
	Ventilator Usage Time in a day	15.9 \pm 7.1 (3.0-24.0)
Treatment of cardiomyopathy	ACEi or ARB	149 (77.6%)
	BB	93 (48.4%)
	CCB	4 (2.1%)
	Diuretics	52 (27.1%)
	Digitalis	31 (16.2%)
	NaBradine hydrochloride	2 (1.0%)
	Antiplatelet	3 (1.6%)
	Antiarrhythmic agent	15 (7.8%)
	No agent for cardiomyopathy	33 (17.2%)
	Ejection fraction (%)	44.8 \pm 14.4 (13.0-78.0)
Nutritional status	Implantable cardioverter-defibrillators	2 (1.0%)
	Oral feeding	162 (84.8%)
	Tube feeding	21 (11.0%)
	Oral + Tube feeding	8 (4.2%)
	Recommended caloric intake	1279.4 \pm 200.1 (825.0-1870.0)

Figure 1. Age distribution of included patients.



Conclusion

There are currently DMD patients of various age groups living in Korea, and their medical conditions vary. Comprehensive management covering diverse fields such as cardiac, respiratory, and nutritional care should be provided to them.