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Cardiac Management of Adult Duchenne Muscular Dystrophy in Korea

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INTRODUCTION

Duchenne muscular dystrophy (DMD) is an X-linked disorder involving the absence of the sarcolemmal protein dystrophin.1 The clinical manifestations of DMD include skeletal muscle degeneration, respiratory insufficiency, and progressive cardiac dysfunction.2 Recent advancements in contemporary cardiopulmonary therapies—including ventilator support—have increased the survival of patients with DMD.3 However, DMD-associated cardiomyopathy remains the leading cause of death.

METHODS

We retrospectively reviewed the charts of all adult DMD patients) who were treated between 2021 and 2022 at a single tertiary referral hospital. A total of 168 patients were enrolled in the study. We excluded patients diagnosed with Becker muscular dystrophy and those with missing transthoracic echocardiography. Mutations within the dystrophin gene were identified using one or more of the following methods: polymerase chain reaction, Southern blotting, DMD gene sequencing, and/or genomic hybridisation array, depending on the technology that was available at the time of diagnosis.

RESULTS

The EF results for different age groups were as follows: for those aged 20 or older but less than 25, EF was 47.4 ± 13.5%; for those aged 25 or older but less than 30, EF was 45.4 ± 14.4%; for those aged 30 or older but less than 35, EF was 40.0 ± 14.2%; for those aged 35 or older but less than 40, EF was 40.7 ± 13.0%; and for those aged 40 or older, EF was 39.6 ± 15.8%. There were no statistically significant differences between the groups. (Figure 1). Out of the 168 patients, 30 individuals (17.9%) did not take medications for cardiomyopathy. In case of using cardiac medication, multiple types of medication were used concurrently. Among those using cardiac medication, multiple types of medication were used simultaneously. Beta-blockers were the most commonly used, with 85 (50.6%) of patients using them. Additionally, ACE inhibitors were used by 59 (35.1%), ARBs (Angiotensin Receptor Blockers) by 64 (38.1%), CCBs (Calcium Channel Blockers) by 3 (1.8%), diuretics by 37 (22.0%), ARNI (Angiotensin Receptor Neprilysin Inhibitor) by 7 (4.2%), antiarrhythmic agents by 3 (1.8%), digitalis by 28 (16.7%), aspirin by 16 (9.5%), clopidogrel by 1 (0.6%), ivabradine by 2 (1.2%), and levocarnitine by 4 (2.4%) of patients (Table 1). Three patients (1.8%) had an Implantable Cardioverter-Defibrillator.

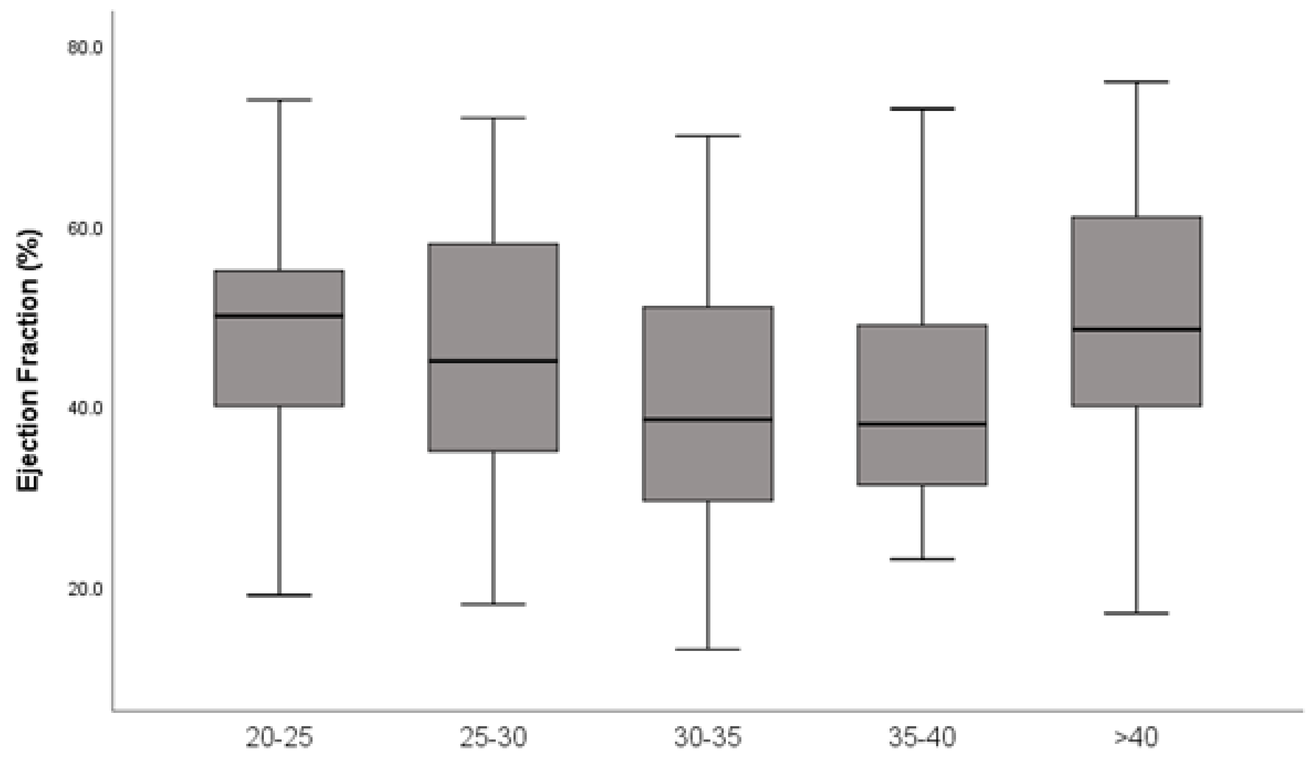

Figure 1. Ejection Fraction by Age Group in patient with DMD

Cardiomyopathy treatment	No.	Percentage (%)
B-blocker	85	50.6%
ACE inhibitor	59	35.1%
ARBs	64	38.1%
CCBs	3	1.8%
Diuretics	37	22.0%
ARNI	7	4.2%
Antiarrhythmic agents	3	1.8%
Digitalis	28	16.7%
Aspirin	16	9.5%
Clopidogrel	1	0.6%
Ivabradine	2	1.2%
Levocarnitine	4	2.4%
No agents	30	17.9%

Table 1. Use of Cardiomyopathy Medications for DMD patients

CONCLUSION

This study highlights the complexities of managing DMD-associated cardiomyopathy and underscores the need for ongoing research to optimize cardiac care in DMD, including the exploration of new therapeutic avenues and strategies to enhance medication adherence and access.