



A PARENT-FOCUSED SURVEY TO ASSESS THE EFFECTS OF THE COVID-19 PANDEMIC ON PATIENTS WITH DUCHENNE MUSCULAR DYSTROPHY

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Abstract: The aim of this study was to evaluate the effect of the Coronavirus Disease (COVID-19) on the use of health and rehabilitation services, physical and general health conditions of individuals with Duchenne Muscular Dystrophy (DMD) from the perspective of parents. A questionnaire consisting of 35 questions was created by the research team. The data were obtained from the parents/primary caregivers of individuals whose diagnoses were confirmed by genetic testing and accessed online via the DMD Families Association. It was reported that 5.5% of the 272 individuals with DMD (age: 10.43±6.42 years) included in the study were infected with COVID-19. It was observed that the major problems encountered during the pandemic were obtaining health services (81%) and stress management (42%). More than half of the individuals were found to be adversely affected in general health (52%), mobility (56%), range of motion (54%), mood (66%), and social dialogs (62%). Furthermore, it was determined that 71% of the individuals took a break from the center where they received outpatient physiotherapy for 3 months or more, and only 6% could receive telerehabilitation service and the level of satisfaction was relatively moderate (10-point visual analog scale: 6.3±2.6). The parents reported that the health care utilization and physical-general health status of individuals with DMD were significantly adversely affected during the COVID-19 pandemic. The present study has an exploratory feature regarding the problems faced by individuals with DMD during the COVID-19 pandemic. Therefore, we think that it will guide future studies in preventing the negative effects of situations such as pandemics or natural disasters.

Keywords: Muscular dystrophy, Duchenne, COVID-19 pandemic, Health care utilization

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1. Introduction

Duchenne muscular dystrophy (DMD), an X-linked recessive progressive muscle disorder caused by mutations in the dystrophin gene, is one of the most common childhood neuromuscular diseases. The clinical course of DMD includes progressive weakness, gait abnormalities, loss of ambulation and motor skills, and, ultimately, progressive respiratory and cardiac failure (Mosqueira et al., 2013; Birnkrant et al., 2018). Although there is still no curative treatment, physiotherapy and corticosteroid treatment remain the mainstays of DMD treatment (Birnkrant et al., 2018).

Individuals with DMD are considered to be at very high risk of severe complications during and after COVID-19 infection due to their reduced respiratory function, cardiomyopathy, immunosuppression from long-term corticosteroid treatment, and obesity (Veerapandiyam et al., 2020). All patients, including DMD, were advised to follow various protective measures, such as self-isolating, not going out of the home, avoiding mixing with others, and social distance, to protect them from contracting

COVID-19 infection (Veerapandiyam et al., 2020). In addition, lockdown and travel restrictions were implemented in many countries. Although vaccination programs are currently underway in many countries, it is still recommended to comply with protective measures since herd immunity cannot be achieved. As a result, the pandemic has resulted in the disruption of healthcare services including physiotherapy as well as schools and social life. This situation has particularly affected children with disabilities, such as DMD, who need help in their daily lives, close contact with caregivers, and regular physical therapy (Matsumura et al., 2021; Nishizawa and Nakamura, 2021). Protective measures and their consequences in individuals with DMD may deteriorate the existing disease, increase caregiver burden, and decrease quality of life. Recent studies indicated that the pandemic adversely affected the frequency of exercise, motor function, functional level, and mental health in individuals with DMD (Matsumura et al., 2021; Sobierajska-Rek et al., 2021a; Sobierajska-Rek et al., 2021b).



It may be helpful to consider that the pandemic may present different challenges and needs for DMD patients and their parents in different countries. Therefore, the primary aim of the present study was to determine parent perceptions of the effects of the COVID-19 pandemic on the physical and general health status of individuals with DMD in our study population. A secondary aim was to determine access to physiotherapy and rehabilitation and telerehabilitation services during the pandemic.

2. Material and Methods

The study was completed between September 2021 and November 2021. The population of this study consisted of individuals with DMD and their parents registered with the Turkish DMD Families Association. Inclusion criteria of the study for parents; having a child with a diagnosis of DMD confirmed by genetic testing, volunteering to participate in the study, and being able to read and write. Individuals with a genetic or neurodevelopmental disorder other than DMD and their parents were excluded from the study. For steroid use of children with DMD, there are no inclusion or exclusion criteria.

An online survey was developed by DMD teams at the Lokman Hekim University Neuromuscular Diseases Application and Research Center on the basis of DMD and COVID-19 related research (Saeed et al., 2020; Karatekin et al., 2021; Nishizawa and Nakamura, 2021; Sancho et al., 2021; Sobierajska-Rek et al., 2021a). We invited parents of individuals with DMD to review the survey and to receive feedback. Additionally, the Director and Scientific Advisory Board of the DMD Families Association reviewed the survey and provided feedback prior to distributing it. The survey asked to be completed by parents included the questions about demographics, COVID-19 infection history, period of stays at home and absence from physiotherapy, challenges encountered by individuals with DMD, and changes in their general health, physical and psychological status during the pandemic (See appendix). The survey, which also includes functional level assessments, was completed through a web link connected to a centralized Google Forms database.

2.1. Functional Level Assessment

While the Vignos Scale with an ordinal classification between 1 and 10 was used to determine the functional level of the lower extremity, the Brooke Scale with an ordinal classification between 1 and 6 was used to determine the functional level of the upper extremity (Vignos and Archibald, 1960; Hiller and Wade, 1992).

2.2. Statistical Analysis

Characteristics of individuals with DMD and their parents were described by using mean (standard deviation) for the numerical variables and by using frequency (%) or median (range) for categorical variables. Data on the

responses to each question were summarized as means, frequencies, and bar graphs, as appropriate. The relationship of age with ordinal variables and non-normally distributed numerical variables was examined with the Spearman correlation test. The statistical analyses were conducted with IBM SPSS Statistics 26.0 (SPSS Inc, Chicago, IL, USA) with the alpha equal to 0.05 (Barton and Peat, 2014).

3. Results

Three hundred and twenty parents who had children with DMD and residing in Turkey responded to the survey (Table 1). Forty-eight parents were excluded from the study due to data duplication (n=41) or no diagnosis of DMD in their children (n=7). Finally, 272 boys with DMD and their parents were included in the study (See flow diagram of study participants: online supporting information).

Table 1. Characteristics of individuals with DMD and their parents

Individuals with DMD, n=272	
Age (years)	10.43 ± 6.42
Diagnostic age (years)	3.29 ± 2.19
Disease duration (years)	7.12 ± 5.55
Brooke Scale median (range)	2 (1-6)
Vignos Scale median (range)	4 (1-10)
History of COVID-19 infection n (%)	15 (5.51)
Respondents, n=272	
Mother n (%)	166 (61)
Father n (%)	89 (33)
Other caregivers n (%)	17 (6)
History of COVID-19 infection n (%)	62 (23)

Most of the respondents (n=166, 61%) were mothers. The mean age and disease duration of the individual with DMD are 10.4 and 7.1 years, respectively. Fifteen of the children (5.5%) and 62 (23%) of the parents had a history of COVID-19 infection. The clinical and demographic characteristics of the individual with DMD are given in Table 1.

More than half of the parents (n=145, 53%) stated that their children stayed at home for 2 months or more, either compulsorily or voluntarily during the pandemic. Most of the parents (n=193, 71%) reported that their children stayed away from the center where they received outpatient physiotherapy for 3 months or more during the pandemic. The most common reason for not going to outpatient physiotherapy centers was fear of contracting COVID-19 infection. While 56% of the parents could not reach any physiotherapy services, 36% continued their routine physiotherapy home program and 6% received physiotherapy services through telerehabilitation during the stay-at-home period (Table 2).

Table 2. Results about stay at home period and physiotherapy service during the pandemic

	n (%)	
The longest period your child stays at home, either compulsorily or voluntarily during the pandemic	A few weeks	83 (31%)
	About 1 month	44 (16%)
	About 2 months	23 (9%)
	About 3 months	28 (10%)
	More than 3 months less than 6 months	42 (15%)
	More than 6 months	52 (19%)
Total time off from the center (such as a special education and rehabilitation center) where you receive outpatient physiotherapy during the pandemic period	about 1-2 months	80 (29%)
	about 3 months	77 (28%)
	More than 6 months Less than 12 months	55 (20%)
	More than 12 months Less than 18 months	26 (10%)
	more than 18 months	34 (13%)
Conditions that disrupt going to outpatient physiotherapy centers	Fear of contracting COVID-19	185 (68%)
	Transportation problems	35 (13%)
	Health problems	47 (17%)
	The closure of outpatient centers	132 (49%)
Physiotherapy service during your stay at home, either compulsorily or voluntarily	Did not receive service	153 (56%)
	Home program	97 (36%)
	Home visiting by a physical therapist	5 (2%)
	Telerehabilitation	16 (6%)

Parents who reported that they had participated in a telerehabilitation session reported a mean satisfaction rating of 6.3 (0= not satisfied; 10= completely satisfied). More than of parents reported that telerehabilitation sessions would be better if there was technological device support and if there were longer and more frequent session time. While half of the parents who had

experienced both in-person and telerehabilitation session preferred in-person sessions, the other half preferred both sessions, none of them preferred telerehabilitation. Most parents reported interruptions in healthcare delivery with reporting canceled or rescheduled healthcare visits. The majority of these visits were physiotherapy appointments (Table 3).

Table 3. Results about telerehabilitation and changed or cancelled healthcare appointments during the pandemic

	n (%)	
Receiving telerehabilitation services	16 (6%)	
Actions to improve telerehabilitation services	Providing device support such as computer / tablet / phone	9 (56%)
	Providing better and faster internet service	5 (31%)
	Increasing the quality of voice and video calls	6 (38%)
	Longer and more frequent service time	9 (56%)
Preferred type of healthcare service	Telerehabilitation	0 (0%)
	Face to Face	8 (50%)
	Both of them	8 (50%)
Rescheduling or cancellation of your child's doctor/physiotherapist appointments due to the pandemic	Yes	220 (81%)
	No	52 (19%)
Changed or canceled appointments	Neurology	95 (43%)
	Physiotherapy	178 (81%)
	Home health care	11 (5%)
	Psychiatry	31(14%)
	Other	5 (2%)

Although there was a negative, lower moderate correlation between age and changes in mobility, there was a weak correlation between age and changes in muscle disease, general health, range of motion, and mood. In addition, there was also a positive correlation between the age and period of stays at home and absence from physiotherapy (Table 4).

The most frequently reported challenges encountered by individuals with DMD during the pandemic were obtaining healthcare services, stress management, feeling alone, and financial issues (Figure 1). Other challenges reported by parents included caregiving and obtaining essentials (medications, groceries, etc). More than half of the parents reported that their

children’s muscle disease (57%), general health (52%), mobility (56%), frequency of exercise (68%), mobility (56%), range of motion (54%), mood (66%), and social dialog (62%) were worse during the pandemic compared before (Figure 2).

Table 4. Relationship between the age and other variables

	Age	
	r	P
Vignos Scale	0.788	<0.001
Brooke Scale	0.659	<0.001
Muscle disease	-0.227	<0.001
General health	-0.241	<0.001
Frequency of exercise	-0.073	0.232
Mobility	-0.305	<0.001
Range of motion	-0.293	<0.001
Mood	-0.147	0.016
Social dialog	-0.094	0.124
Period of stays at home	0.292	<0.001
Period of absence from physiotherapy	0.305	<0.001

4. Discussion and Conclusion

The results of this study showed that the families and caregivers of the children diagnosed with DMD who participated in the survey caused the COVID-19 Pandemic to cause disruptions in the health services they receive and interrupt their routine rehabilitation services. Caregivers stated that they and their children had difficulties due to social distance and pandemic restrictions. According to the results of our survey conducted in the DMD group, it was reported that social communication and mental status deteriorated by 59.7% and 62.8%, respectively. In another study, anxiety, stress, and depression symptoms were found in caregivers of children with special needs in India (Ping et al., 2020). In a study conducted in China, anxiety was reported as one of the two most frequently reported problems during the pandemic. The fear of catching COVID-19 and the protective approach of families have closely affected the DMD group, which has a high risk of COVID-19 transmission and infection. Suspension of the activities of the centers where the DMD group receives special education and rehabilitation routinely, and the outpatient hospitals not accepting patients to the treatment units except for emergencies, adversely affected the physical and functional capacities and mental states of the children. A study conducted in France reported that 77% of children’s medical consultations were canceled during quarantine (Cacioppo et al., 2021). The life habits of children with DMD have also changed due to environmental restrictions applied within the scope of health policies. In addition, with the closure of schools and rehabilitation centers, the effects on fear of COVID-19 have decreased due to the possibility of infection to their children and themselves.

In individuals with DMD, a negative correlation was found between age and general health, physical capacity (level of mobility, range of motion), and mental state, while a positive correlation was found between age and functional status, length of stay at home, and the duration of not receiving physiotherapy disease and worsening of symptoms with increasing age support these results. These results are in line with the progressive nature of DMD with increasing age (Falzarano et al., 2015).

Goldschmidt et al. (2020) stated that children and their families should be supported socially and emotionally with technology-based approaches. In the Pandemic period, when the benefits of telemedicine are comprehensively discussed, there is a need to bring patient satisfaction to the fore and integrate multidisciplinary approaches into the system in the process of continuing calls for action to increase telehealth. Being a rare and progressive disease, DMD is a potentially higher risk for poor outcomes in patients being infected with COVID-19 due to physical disabilities, age, mobility status, and cardiopulmonary involvement.

It should not be forgotten that exercise training can improve and protect the functional status of DMD patients, and it is of great importance for this group to follow and update the exercises determined by the telerehabilitation program to be given considering the risks. In cases where face-to-face health services cannot be provided due to the COVID-19 pandemic and similar reasons, the pre-configuration and availability of remote services will prevent disruption of the treatment of this group. Tele-rehabilitation will be indispensable in today’s treatments, as it helps to reach more patients, remove the barrier of access to treatment, improve quality of life and provide more frequent evaluations. There is an increasing number of studies showing that telerehabilitation can be as effective as face-to-face treatments in pediatric rehabilitation (Camden et al., 2020). In addition, reducing the transportation fee to be spent for telerehabilitation health care services, spending the time to be spent for treatment and control more efficiently, in accordance with social distance, will protect patients and their families both during the pandemic period and in similar situations, preventing them from disrupting the control and treatment.

In our study, 16 (%6) of children with DMD received telerehabilitation service and their telerehabilitation satisfaction was relatively insufficient (6.3 out of 10), which indicates the lack in this regard in Turkey. The results in our study are not comparable with other studies due to insufficient participation of children in Telerehabilitation. Telerehabilitation satisfaction in the absence of face-to-face contact with DMD boys and their families will guide further pandemics that may occur in the possible future of increasing telehealth. For this reason, the telerehabilitation service should be addressed and the quality of service should be increased by working on its deficiencies. Our survey was planned to show clinicians and researchers the effects of the COVID-

19 Pandemic on the lives of individuals with DMD and their caregivers. Limitations of this study the general health status of children with the questionnaire is obtained from the subjective information given by the

caregivers. In addition, there is no study in which the data obtained as a result of our survey can be compared with the pre-pandemic data.

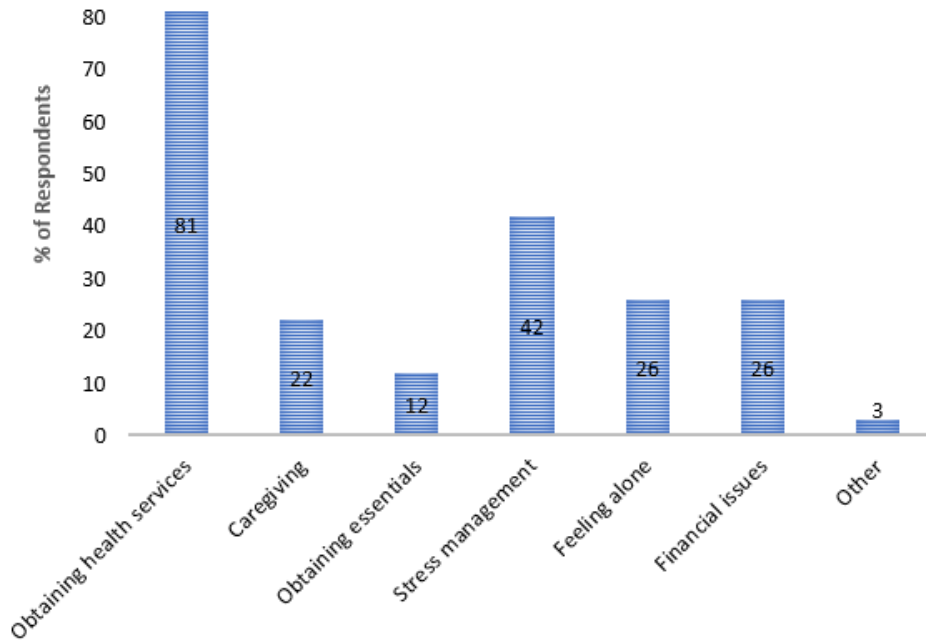


Figure 1. Challenges encountered by individuals with DMD during the COVID-19 Pandemic.

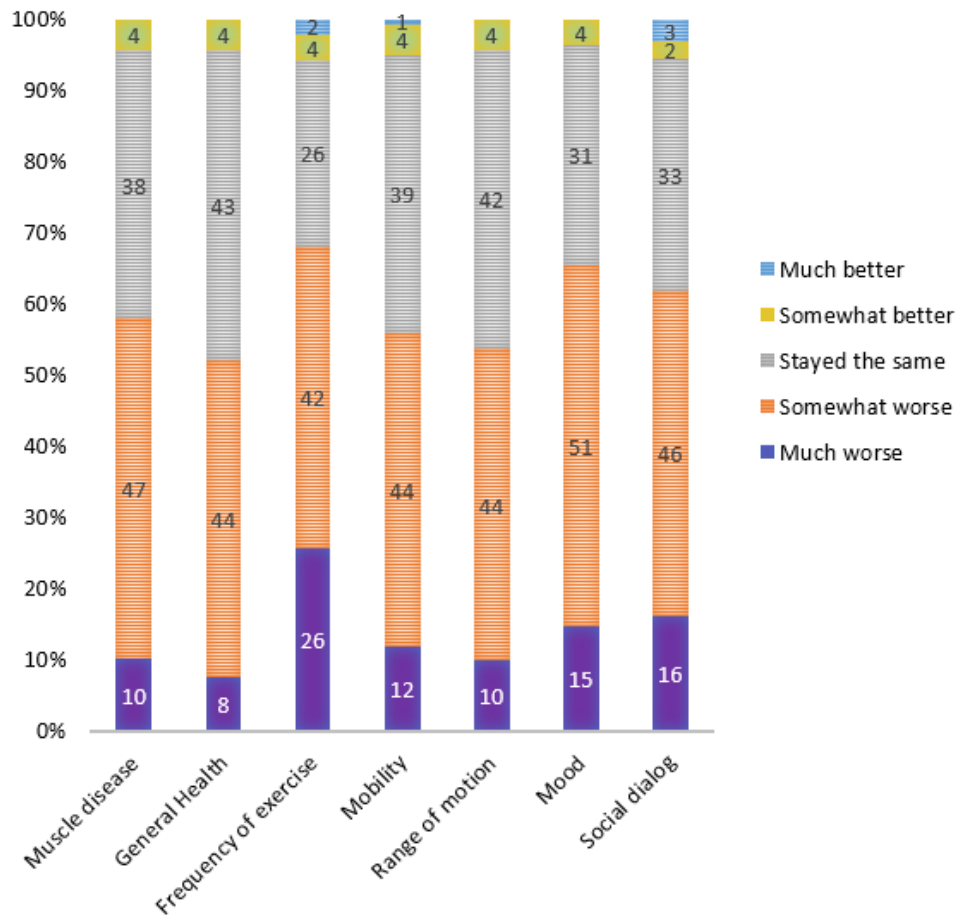


Figure 2. Changes in general health, physical and psychological status of individuals with DMD during the COVID-19 pandemic

Author Contributions

All authors contributed to the study conception and design. The first draft of the manuscript was written by A.R.T. and all authors commented on previous versions of the manuscript. All authors read and approved the final manuscript.

Conflict of Interest

The author declared that there is no conflict of interest.

Ethical Approval/Informed Consent

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. The study was approved by the Non-interventional Clinical Research Ethics Committee of Lokman Hekim University (September 13, 2021, GO: 2021102).

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