



A patient-focused survey to assess the effects of the COVID-19 pandemic and social guidelines on people with muscular dystrophy

Katy Eichinger PT, PhD¹ | Leann Lewis MS¹ | Nuran Dilek MS¹ |
Kiley Higgs MS² | Michaela Walker MPH² | David Palmer³ | John M. Cooley³ |
Nicholas Johnson, MD⁴  | Rabi Tawil, MD¹ | Jeffrey Statland, MD² 

¹Department of Neurology, University of Rochester, Rochester, New York, USA

²Department of Neurology, University of Kansas Medical Center, Kansas City, Kansas, USA

³Patient collaborator, University of Kansas Medical Center, Kansas City, Kansas, USA

⁴Department of Neurology, Virginia Commonwealth University, Richmond, Virginia, USA

Correspondence

Katy Eichinger, Department of Neurology,
University of Rochester, 601 Elmwood Ave.
Box 673, Rochester, NY 14642, USA.
Email: katy_eichinger@urmc.rochester.edu

Abstract

Introduction/Aims: In this study, we examined the social and health impacts of the coronavirus disease 2019 (COVID-19) pandemic and social guidelines on people with muscular dystrophies.

Methods: A prospective de-identified electronic survey was distributed to adults with self-reported facioscapulohumeral muscular dystrophy (FSHD), myotonic dystrophy (DM), and limb-girdle muscular dystrophy (LGMD) enrolled in national registries or with patient advocacy groups.

The COVID-19 Impact Survey was developed by muscular dystrophy experts in association with patient collaborators and advocacy groups. The Perceived Stress Scale was used to measure perceived stress.

Results: Respondents (n = 774: 56% FSHD; 35% DM, and 9% LGMD) were mostly women and middle-aged (range 19–87 y). Rates of COVID-19 infections were low (<1%), compliance with local social distancing guidelines and policies high (98%). Major challenges reported during the pandemic included: obtaining treatment (40%), managing stress (37%), social distancing (36%), and obtaining essentials (34%). The majority reported a slight worsening in their disease state. Respondents reported moderate stress levels (stress score = 15.4; range = 0–35), with higher stress levels reported by women and those under age 30 y. Three-quarters of participants who participated in telemedicine visits were satisfied with the encounters; however, most reported a preference for in-person visits.

Discussion: People with muscular dystrophy reported moderate stress and challenges during the COVID-19 pandemic. Interventions such as exercise and stress-coping strategies, including strategies specific to women or individuals <30 y, may be

Abbreviations: COVID-19, coronavirus disease 2019; FSHD, facioscapulohumeral muscular dystrophy; LGMD, limb-girdle muscular dystrophy; MD, muscular dystrophy; DM, myotonic dystrophy; PSS, Perceived Stress Scale.

Katy Eichinger and Leann Lewis contributed equally to this study.

important. Further investigation is needed into the role of telemedicine in the care of individuals with muscular dystrophy.

KEYWORDS

COVID-19, muscular dystrophy, pandemic, perceived stress scale, telemedicine

1 | INTRODUCTION

The coronavirus disease 2019 (COVID-19) pandemic and the related national and local social guidelines have required unprecedented changes in social conventions worldwide; most guidelines are driven by the need to control the spread of the pandemic. Many of our basic principles of social gathering and trade have come under scrutiny, and most COVID-19 guidelines have focused on social isolation (stay-at-home orders) or social distancing.^{1,2} These guidelines have also limited access to medical care or caused changes to care delivery strategies, including a rapid transition from face-to-face encounters with healthcare providers to the use of telehealth or telephone visits.³ In addition, certain populations, like individuals over the age of 60 y, with cardiac or respiratory disease were encouraged to avoid public spaces.⁴ Certain types of muscular dystrophy (MD) can be associated with cardiopulmonary complications, thus putting these patients at higher risk for COVID-19.

The standard of care for MD includes care provided in multidisciplinary clinics and many of these clinics “paused” patient care at the beginning of the pandemic. Standard interventions such as physical therapy may not be available. Individuals with MD may be homebound secondary to mobility issues, and may require assistance from caregivers or home health providers, which may be limited. The effect the COVID-19 pandemic and associated guidelines on people with MD is currently unknown.

We conducted a survey of people with MD to determine the impact of COVID-19 and the associated social guidelines on them and to examine individuals' perceived stress during the early stages of the pandemic in order to help guide future health interventions to improve care. A secondary goal is to determine the response to healthcare delivery adaptations like telemedicine, and the impact on ongoing research programs.

2 | METHODS

We conducted a prospective de-identified electronic survey from May 8, 2020, to May 28, 2020, of people with MD residing in the United States. Participants completed the survey through a Web-link connected to a centralized REDCap database.⁵ The study was approved by the University of Rochester Medical Center Institutional Review Board, and exempted from HIPAA consent requirements, as all surveys were de-identified. Participants had self-reported facioscapulohumeral dystrophy (FSHD), myotonic dystrophy (DM), or limb girdle muscular dystrophy (LGMD), and were registered with the following national registries

or patient advocacy groups: C3 Patient Registry – Coalition to Cure Calpain 3 (curecalpain3.org), the Dysferlin Registry – Jain Foundation (jain-foundation.org), the FSHD Society (fshdsociety.org), the Myotonic Dystrophy Foundation (myotonic.org), the National Registry for Myotonic Dystrophy & Facioscapulohumeral Dystrophy housed at the University of Rochester Medical Center (dystrophyregistry.org), and the Speak Foundation (thespeakfoundation.com). An institutional review board-approved information letter with an embedded link to the survey was sent to registry participants who had agreed to be contacted by email or conventional mail, or posted as notifications on advocacy websites. The survey link was also provided to registries who distributed it by posting on a website, including in a newsletter, sending a batch email, or putting out a press release. While the exact number who received the link is unknown, the registries have approximately 7000 registrants total, including affected individuals, their family members, or healthcare providers. To minimize the possibility of duplicate surveys of individuals registered at multiple registries, the first question was a stop-end question asking respondents if they had completed the survey before.

2.1 | Measures

The COVID-19 Impact Survey was developed by muscular dystrophy teams at the University of Rochester Medical Center (R.T., K.E., L.L.), University of Kansas Medical Center (J.S., K.H., M.W.), and Virginia Commonwealth University (N.J.). We invited individuals with muscular dystrophy to review the survey, and we received feedback from three individuals with FSHD and two individuals with DM. Additionally, the Director of Research and Patient Engagement at the FSH Society, the Scientific Director of the Coalition to Cure Calpain 3, the Myotonic Dystrophy Foundation Scientific Advisory Board provided feedback, and advocacy representatives reviewed the survey prior to distributing it. Survey categories included: demographics and disease history, COVID-19 medical history, and the impact of the pandemic and associated social guidelines on disease-specific topics such as exercise, pain, stress, and medical care. The survey also explored experience with telemedicine visits and research study participation. (See Supporting Information Materials, which are available online).

The Perceived Stress Scale (PSS) is a validated 10-item scale that assesses the amount that situations in a person's life are perceived as stressful.⁶⁻⁸ Each item is scored 0–4 with a total score of 0–40 (higher scores indicating increased stress). A stress score of 0–13 is considered low stress, 14–26 moderate stress, and a score of 27–40 indicates high stress.^{8,9}

2.2 | Statistical analysis

Data were summarized by responses to each question: frequencies, means, and SDs. Associations between discrete or continuous data elements were explored using Pearson correlations.¹⁰ Differences in perceived stress between groups based on clinical diagnosis, gender, age categories, or ambulatory status was performed by analysis of variance using Tukey's method for multiple comparisons, or T-test. Where there were differences by diagnosis, an analysis of covariance was performed to adjust for potential demographic differences. Statistical analysis was performed using SPSS version 26 (IBM, Armonk, NY). For this exploratory survey, all statistical testing was two-sided and a *P*-value <.05 is significant.

3 | RESULTS

3.1 | Participants

Seven hundred and seventy-four adults with MD residing in the United States responded to the survey (Table 1). Respondents were approximately equally divided between the sexes, with the exception of those with DM, who were predominantly women. The majority of the participants reported genetic confirmation and most individuals reported living with other people. Approximately a third reported using a wheelchair either part-time or full-time.

3.2 | Social strategies

Social distancing (95%), avoiding crowds larger than 10 (86%), and wearing masks (84%) were reported by the majority of participants as social strategies. Participants reported stay-at-home orders less frequently (78%). Ninety-eight percent of the participants reported

adhering to the local social distancing recommendations that were in place in their state. The most frequently reported challenges during the COVID-19 pandemic were obtaining treatment, managing stress, social distancing, and obtaining essentials (groceries, medications, etc) (Figure 1A). Other challenges reported by respondents included lack of access to exercise, lack of access to mental health services, isolation, and change in work (ie, working from home, working more/less hours). Figure 1B summarizes the challenges experienced due to social distancing with the most frequently reported being loneliness and performing daily tasks.

3.3 | Perceived stress

The mean PSS score was 15.4, indicating moderate stress. Individuals with DM, women, and individuals younger than 30 y of age reported significantly higher levels of stress (Table 2). However, when adjusted for sex, the difference in stress scores between disease conditions did not remain. There was a low, significant, negative correlation between lower PSS scores and ability to manage stress ($r = -0.47$; $P < .0001$). There was also a low, significant correlation between higher PSS scores and individuals who reported higher pain levels ($r = 0.30$; $P < .0001$).

The majority of participants (88%) reported that they have been able to manage their stress and have done so most frequently by resting and sleeping, exercising, and meditating. Other ways respondents reported managing stress included religion/faith, family and friend interactions, and hobbies (Figure 2).

3.4 | Impact on health and access to medical care

Over half of the individuals reported that their muscle disease did not change while a third reported a slight worsening of their muscle

TABLE 1 Patient characteristics

Characteristic	FSHD	DM	LGMD	Overall
Number	434	271	69	774
Mean age (y, range, SD)	55.6 (21–87; 13.6)	53.7 (21–81; 12.3)	45.0 (19–78; 16.0)	54 (19–87; 13.7)
Female (n, %)	183 (42.1%)	235 (86.7%)	41 (59.4%)	459 (60.0%)
Genetically confirmed (n, %) ^a	361 (83.6%)	243 (90.0%)	69 (100%)	673 (87.3%)
Living situation (n, %) ^b				
Alone	70 (16.6%)	34 (12.7%)	9 (13.0%)	113 (14.9%)
With others	351(83.3%)	234 (87.3%)	60 (87.0%)	645 (85.1%)
Ambulatory status (n,%) ^c				
Uses wheelchair full-time	84 (19.4%)	6 (0.02%)	23 (33.3%)	113 (14.7%)
Uses wheelchair part-time	84 (19.4%)	53 (19.6%)	17 (24.6%)	154 (20.0%)
Does not use wheelchair	264 (61.1%)	211 (78.1%)	(42.0%)	504 (65.5%)

^an = 432 FSHD, n = 268 DM, total n = 769.

^bn = 421 FSHD, n = 268 DM, Total n = 758.

^cn = 432 FSHD, n = 270 DM, total n = 771.

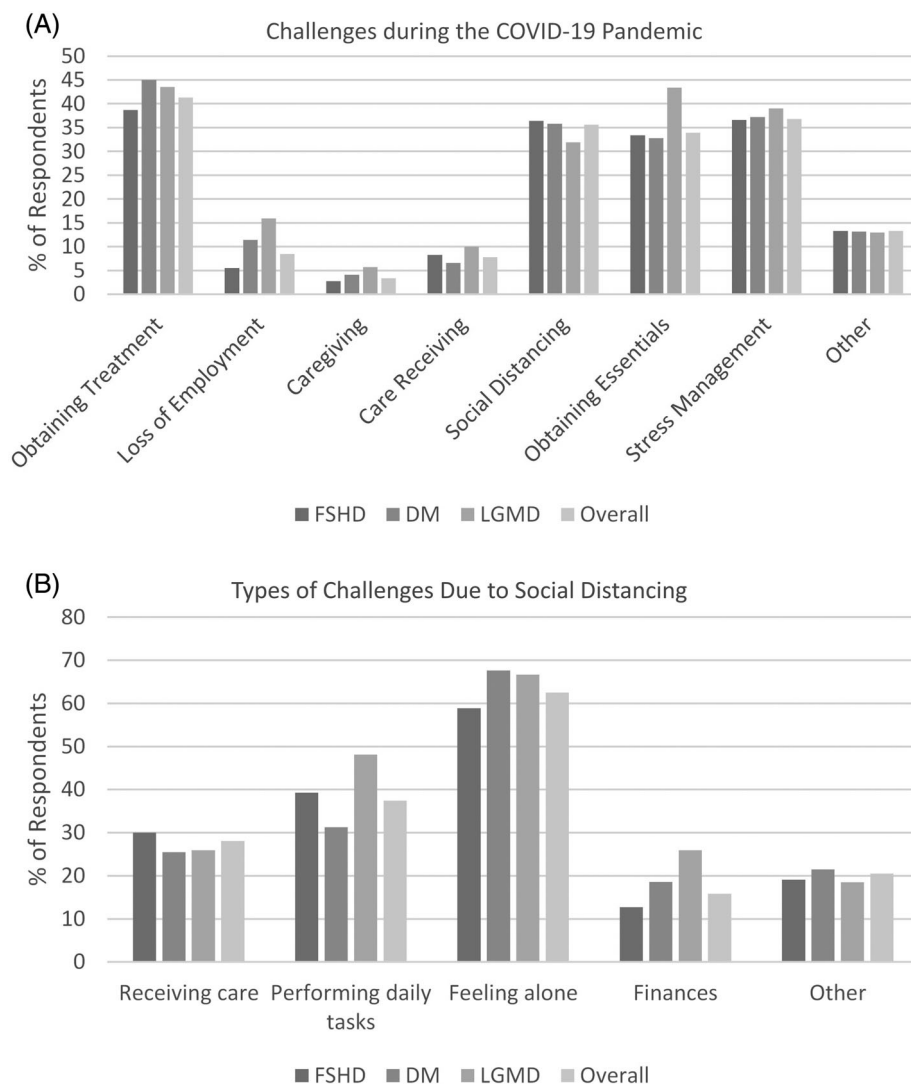


FIGURE 1 Challenges faced during the COVID-19 pandemic. A, The most common challenges identified were consistent across muscular dystrophies, and included obtaining treatment, social distancing, obtaining essentials, and stress management. B, Looking closer at challenges related to social distancing the most common were feeling alone and performing daily tasks

disease. Forty-three percent of individuals reported that their exercise had decreased. The majority of individuals reported pain with 47% reporting that their pain had worsened during the pandemic.

A small percentage of individuals (2.2%) reported that their condition had improved during the pandemic. Of these individuals, 82% reported exercising with 71% reporting increasing their exercise; 67% reported improvement in their pain; and all reported being able to manage their stress. In contrast, a similar percentage reported that their muscle disease was much worse. Of these individuals, 61% reported exercising with half reporting a decrease in exercise; the majority of these individuals reported that their pain had increased, and only 44% reported that they were able to manage their stress.

Individuals reported interruptions in healthcare delivery with two-thirds reporting canceled or rescheduled healthcare visits. The majority of these visits were primary care appointments, followed by neurology and therapy (physical or occupational therapy). Forty-two percent of participants reported that they had participated in a telemedicine appointment via telephone calls or video conferencing platforms. Participants reported a mean satisfaction rating of 75.7 (0 = not satisfied; 100 = completely satisfied), which was

consistent across platforms and diseases. A small percentage of individuals (10%) reported that telemedicine visits would be better if they were easier to access and use and if there was audio/visual improvement. The majority of individuals (74.7%) who had experienced both in-person and telemedicine healthcare appointments preferred in-person appointments.

Thirteen percent of the participants reported participating in research studies and half of these participants reported that the study had been affected by the pandemic. Participants reported delayed or canceled study visits and/or modifications to study procedures. Despite these challenges, 94% reported that their willingness remained unchanged, or they would be more likely to participate in future research studies.

4 | DISCUSSION

This study revealed that the COVID-19 pandemic and associated guidelines had an impact across social and medical needs of people with MD, and that the vast majority of individuals adhered to local

Addressing issues of stress, pain, and exercise could be a focus for providers when developing mitigation strategies during the COVID-19 pandemic.

Individuals reported access to care as a major challenge due to COVID-19 across all MD diseases. Many healthcare visits were cancelled or rescheduled, and it is not known how the delay in care affected or will affect individuals' overall health. Our survey showed the majority of participants were satisfied with their care via telemedicine; however, a preference for in-person visits was apparent. This suggests a need for better understanding of the preference for in-person visits to help improve telemedicine so that it can serve as an effective and safe substitution while COVID-19 risks are still substantial. In a time where the benefits of telemedicine are being widely discussed, and calls to action for increasing telehealth are underway,³ a better understanding of the drivers of patient satisfaction and different multidisciplinary care delivery models is also needed.

Higher stress levels in individuals with MD may have been anticipated due to physical barriers and potential higher risk for poor outcomes if infected with COVID-19, due to age, mobility status, or cardiopulmonary involvement of MD. In our study, however, we found moderate stress levels, and PSS scores similar to what was reported in a worldwide study looking at the stress of the pandemic in a general population.^{14,15} Also similar, we found higher stress levels in women and those under 30 y of age. One explanation for this difference may be women are more likely than men to disrupt their daily lives by following social distancing guidelines, staying home from work, and being the primary caregivers of children and sick or elderly family members.¹⁶ The US Census Bureau Household Pulse Survey also showed that individuals ages 18 to 29 in the general population have consistently had the highest rates of anxiety and depression symptoms during the pandemic.¹⁷ Younger adults may have less job security and financial worries that may result in greater stress levels than older individuals. Focusing on stress management in women and young individuals would be important when considering future interventions.

Research has become a main priority for the MD community, as transformational gene-based therapies are currently being approved for inherited neuromuscular diseases.¹⁸ The effect of the pandemic on the willingness of people with MD to participate in research was unknown. Despite delayed or canceled study visits, and modified study procedures during the pandemic,^{19,20} study participants have showed great flexibility during this time, and it is encouraging that these experiences have not decreased their likelihood for participating in future research.

There are several limitations to this study. We designed this study in response to the pandemic and therefore we do not have data on how these individuals would have responded to these questions prior to the pandemic. Additionally, we did not collect data on a non-muscular dystrophy sample for comparison and, therefore, could only compare our findings with other reports in the literature. We are also limited by bias that may affect the decision to participate in patient registries or respond to surveys, which may select for more or less affected individuals, individuals without cognitive involvement (DM) or for older individuals who have time to participate in registries. We cannot eliminate the possibility that someone would be in more

than one registry and may have responded to the survey twice; although we included a question to minimize this possibility. Additionally, some of the data collected, such as the PSS score, have no published studies in MD prior to the pandemic to compare against; but the values found in this study are important as it will allow for future comparison as guidelines and responses to the pandemic continue to evolve. Finally, the use of an electronic survey may select for people with the economic resources to have electronic devices and Internet access in their home, and for people comfortable with using the technology.

CONCLUSIONS

Our survey provided an opportunity for individuals with muscular dystrophy to make their voices heard, to contribute to the current research remotely, and ensure clinicians, researchers, and advocacy groups understand the impact of the COVID-19 pandemic and social guidelines on their lives. It also helps us understand their respective muscle diseases in a unique environment and helps us to continue to provide high-level care in this period. Understanding the impact on social and medical aspects of care can guide future interventions, and the approach taken here can serve as a model for other rare, chronic progressive diseases.

CONFLICT OF INTEREST

K.E. serves as a consultant and has received consulting fees from Ionis, Biogen, Acceleron, Fulcrum, and Avidity. N.J. reports grants from NINDS (R01NS104010), grants from NCATS (R21TR003184), grants from CDC (DD19-002), grants from FDA (R01FD006071), grants from Muscular Dystrophy Association, grants from Myotonic Dystrophy Foundation, grants from Coalition to Cure Calpainopathies, grants and personal fees from Dyne, grants and personal fees from AveXis, grants and personal fees from Vertex Pharma, grants and personal fees from Fulcrum Therapeutics, grants and personal fees from ML Bio, grants and personal fees from Sarepta, grants and personal fees from Acceleron, grants from AMO Pharma, outside the submitted work; In addition, NJ has a patent CMTHI with royalties paid, and a patent CCMDHI issued. R.T. serves on advisory boards for Fulcrum Therapeutics, Dyne therapeutics and Arrowhead Pharma. J.S. received grant support from NIH, FSHD Society, Muscular Dystrophy Association, Friends of FSH Research. JS served as a consultant or on the advisory boards for Acceleron, Fulcrum, Dyne, Strongbridge, Sarepta, Avidity, and Genzyme.

L.L., N.D., K.H., M.W., D.P., and J.C. report no disclosures.

ETHICAL PUBLICATION STATEMENT

We confirm that we have read the Journal's position on issues involved in ethical publication and affirm that this report is consistent with those guidelines.

DATA AVAILABILITY STATEMENT

The data that support the findings of this study are available from the corresponding author upon reasonable request.

ORCID

Nicholas Johnson  <https://orcid.org/0000-0002-3917-4257>

Jeffrey Statland  <https://orcid.org/0000-0003-0790-5315>

REFERENCES

- Shatzkes M & Rai K. Introducing the health act: Expanding medicare reimbursement. <https://www.sheppardhealthlaw.com/2020/06/articles/healthcare-law/healthcare-legislation-and-rulemaking/health-act-of-2020>. Accessed July 1, 2020.
- Guner R, Hasanoglu I, Aktas F. COVID-19: prevention and control measures in community. *Turk J Med Sci*. 2020;50:571-577.
- Ohannessian R, Duong TA, Odone A. Global telemedicine implementation and integration within health systems to fight the COVID-19 pandemic: a call to action. *JMIR Public Health Surveill*. 2020;6:e18810.
- Centers for Disease Control and Prevention. Communities, schools, workplaces, and events. <https://www.cdc.gov/coronavirus/2019-ncov/community/index.html>. Accessed June 22, 2020.
- Harris PA, Taylor R, Thielke R, Payne J, Gonzalez N, Conde JG. Research electronic data capture (REDCap): a metadata-driven methodology and workflow process for providing translational research informatics support. *J Biomed Inform*. 2009;42(2):377-381.
- Cohen S, Kamarck T, Mermelstein R. A global measure of perceived stress. *J Health Soc Behav*. 1983;24:385-396.
- Cohen S, Williamson G. Perceived stress in a probability sample of the United States. In: Spacapan S, Oskamp S, eds. *The Social Psychology of Health, Claremont Symposium on Applied Social Psychology*. Newbury Park, CA: Sage Publications, Inc.; 1988.
- Cohen S, Janicki-Deverts D. Who's stressed? Distributions of psychological stress in the United States in probability samples from 1983, 2006 and. *J Appl Soc Psychol*. 2009;2012:1320-1334.
- Cohen, S. Perceived stress scale. Mind Garden <http://www.mindgarden.com/documents/PerceivedStressScale.pdf>. Accessed April 7, 2020.
- Hinkle DE, Wiersma W, Jurs SG. *Applied Statistics for the Behavioral Sciences*. 5th ed. Boston, MA: Houghton Mifflin; 2003.
- Veerapandiyan A, Connolly AM, Finkel RS, et al. Spinal muscular atrophy care in the COVID-19 pandemic era. *Muscle Nerve*. 2020;62:46-49.
- Veerapandiyan A, Wagner KR, Apkon S, et al. The care of patients with Duchenne, Becker, and other muscular dystrophies in the COVID-19 pandemic. *Muscle Nerve*. 2020;62:41-45.
- Stults-Kolehmainen MA, Sinha R. The effects of stress on physical activity and exercise. *Sports Med*. 2014;44:81-121.
- Zheng Z, Peng F, Xu B, et al. Risk factors of critical & mortal COVID-19 cases: a systematic literature review and meta-analysis. *J Infect*. 2020;81(2):e16-e25.
- Gamonal-Limcaoco S, Montero Mateos E, Fernandez J, Roncero C. Anxiety, worry and perceived stress in the world due to the COVID-19 pandemic, march 2020. Preliminary results. *medRxiv*. 2020. <https://doi.org/10.1101/2020.0403.20043992>
- Hamel L, Salganicoff, A. Is there a widening gender gap in coronavirus stress, <https://www.kff.org/coronavirus-policy-watch/is-there-widening-gender-gap-in-coronavirus-stress>. Accessed July 9, 2020.
- Mental Health. <https://www.cdc.gov/nchs/covid19/pulse/mental-health.htm>. Accessed July 9, 2020.
- Kariyawasam D, Alexander IE, Kurian M, Farrar MA. Great expectations: virus-mediated gene therapy in neurological disorders. *J Neurol Neurosurg Psychiatry*. 2020;91(8):849-860.
- Lupkin S. Coronavirus Pandemic Brings Hundreds Of U.S. Clinical Trials To A Halt. <https://www.npr.org/sections/health-shots/2020/04/11/832210606/coronavirus-pandemic-brings-hundreds-of-u-s-clinical-trials-to-a-halt> Accessed July 9, 2020.
- Epilepsy ILA. Suspended studies and virtual lab meetings: How the COVID-19 pandemic is affecting epilepsy researchers. https://www.newswise.com/coronavirus/suspended-studies-and-virtual-lab-meetings-how-the-covid-19-pandemic-is-affecting-epilepsy-researchers/?article_id=734062. Accessed July 9, 2020.

SUPPORTING INFORMATION

Additional supporting information may be found online in the Supporting Information section at the end of this article.

How to cite this article: Eichinger K, Lewis L, Dilek N, et al. A patient-focused survey to assess the effects of the COVID-19 pandemic and social guidelines on people with muscular dystrophy. *Muscle & Nerve*. 2021;64(3):321-327. <https://doi.org/10.1002/mus.27349>