

How to define and enhance diagnostic and assistance pathways in neuromuscular diseases during the COVID-19 pandemic: the concept of network

Guja Astrea¹, Gemma Marinella^{1,2}, Caterina Agosto³, Delia Gagliardi⁴, Marina Grandis⁵, Maria Giuliano⁶, Luisa Politano⁷

¹IRCCS Fondazione Stella Maris, Calambrone (PI), Italy; ²Department of Clinical and Experimental Medicine, University of Pisa, Pisa, Italy; ³Paediatric Palliative Care, Pain Service, Department of Women's and Children's Health, University of Padua, Padua, Italy; ⁴Dino Ferrari Centre, Department of Pathophysiology and Transplantation (DEPT), Neuroscience Section, University of Milan, Milan, Italy; ⁵IRCCS Foundation Ca' Granda Ospedale Maggiore Policlinico, Neurology Unit, Milan, Italy; ⁶Department of Neuroscience, Rehabilitation, Ophthalmology, Maternal and Child Health University of Genova; ⁷IRCCS San Martino, Genoa, Italy; ⁸Regional President of SIMPe Campania (Italian Society of Pediatricians) and National Head of SIMPe for Rare and Chronic Diseases; ⁹Cardiomyology and Medical Genetics, University of Campania, Naples, Italy

Received: October 30, 2021
Accepted: December 12, 2021

Correspondence

Guja Astrea
IRCCS Fondazione Stella Maris, 56127 Calambrone (PI), Italy. E-mail: guja.astrea@fsm.unipi.it

How to cite this article: Astrea G, Marinella G, Agosto C, et al. How to define and enhance diagnostic and assistance pathways in neuromuscular diseases during the COVID-19 pandemic: the concept of network. *Acta Myol* 2021;40:172-176. <https://doi.org/10.36185/2532-1900-060>

© Gaetano Conte Academy - Mediterranean Society of Myology



OPEN ACCESS

This is an open access article distributed in accordance with the CC-BY-NC-ND (Creative Commons Attribution-NonCommercial-NoDerivatives 4.0 International) license. The article can be used by giving appropriate credit and mentioning the license, but only for non-commercial purposes and only in the original version. For further information: <https://creativecommons.org/licenses/by-nc-nd/4.0/deed.en>

The main consequence of the COVID-19 pandemic has been to increase the distance between patients and their doctors and to limit the opportunities to compare experiences and clinical cases in the medical community. Based on this, we adopted a strategy to create networks with the ambition to break down these distances and to unify the process of care and management. Here we report the results and perspectives of our efforts and studies. A summary of the presentations on the topic, held during the webinars organized for macro-areas by the Italian Association of Myology with the aim of raising awareness among “non-expert doctors” who deal with neuromuscular disorders in the era of COVID-19 was collected and here reported. Although the macro-areas responded in different way to the problems of neuromuscular patients in the era of COVID-19, they all have tried to create a network between doctors and opportunity for education and information, with the secondary outcome to have shared process of care and management. Telemedicine, virtual meetings and the strengthening of national and international networks, through research projects, were the nodal and common points. Due to their complexity, neuromuscular diseases had already taught clinicians the importance of multidisciplinary confrontation. COVID-19 has further strengthened the need to create links between clinicians and experts, even of different nationalities, in order to guarantee to patients the best possible care, but above all, access and continuity of care even in critical periods. Adequate answers have been given to these problems, though there is still a lot to improve.

Key words: neuromuscular disorders, COVID-19 pandemic, telemedicine, networks

Introduction

The impact of coronavirus disease 19 (COVID-19) pandemic in the care of many diseases, and in particular rare, chronic and disabling diseases such as Neuromuscular Disorders (NMDs) has been significant.

National health care services underwent a radical reorganization, with in-person consultations being postponed, and not considered urgent treatments delayed or canceled. In addition, there was a rapid implementation of remote approaches to patients¹.

Therefore, patients with NMDs not having guarantees of care and treatment as before the pandemic were at greater risk of developing severe acute respiratory syndrome-coronavirus-2 (SARS-CoV-2) infection² and experienced deeper distress than to other patients, and prolonged home isolation¹.

A multicentric national survey³ promoted by the Italian Association of Myology (AIM) showed since the first period of the pandemic, a significant malfunction of clinical and support services for patients affected by NMDs at national level, with the following outcomes. First, 40% of surveyed tertiary neuromuscular centers reported a reduction in outpatient visit and examinations, while 22% postponed in-hospital administration of therapies and the vast majority (93%) reduced or suspended rehabilitative services and on-site outpatient visits. Second, the possible worsening of all NMD conditions due to the indirect consequences of the pandemic SARS-CoV-2 infection, as well as the need to stop or significantly restrict the access to clinical trials, was also reported³.

Because of these critical issues, the community of NMDs medical experts tried to reconsider the gold standard of excellence in disease management, evaluating the risk/benefit ratio of specific interventions (pharmacological and rehabilitative), with respect to the possibility of exposure to the disease. They also re-thought the clinical trial procedures to ensure remote participation in all phases of the study, from enrollment to drug administration and follow-up⁴.

This effort was also made in Italy with the aim of creating an approach shared by all Italian third-level centers and facilitating an early diagnosis also at local level.

Thus, the AIM Board proposed a series of webinars aimed at raising awareness among “less experienced” doctors to create a network that could support patients, despite the lack of medical care and the isolation imposed by the COVID-19 pandemic.

Here we report the Italian experience derived from sharing a “new approach to care” during these webinars divided into two macro-areas of our Country (North-Center and Center-South Italy).

Methods

The experience on the management of the neuromuscular patient during the COVID-19 pandemic, shared through lectures presented in the webinars and entitled “The management of the patient with neuromuscular disease in the pandemic period: from telemedicine to the vaccine experience” is proposed below by comparing, for simplicity, two national macro-areas.

Results

North-Central Italy

In the North-Center, the first Italian macro-area that faced the spread of COVID-19 pandemic, the main strategy adopted was to create networks to reduce the distances imposed by the pandemic and to standardize the care and management process. This was possible by defining three types of networks, networks between stakeholders, regional networks and national/international networks. To improve the network between doctor and patient, face-to-face visits were replaced by telemedicine appointments. This modality cannot replace the direct neuromuscular examination but has the advantage of monitoring the patient’s clinical status through anamnestic collection and the tests performed.

A rehabilitation experience using telemedicine was also reported⁵. Notably, a 28-year-old man with Charcot-Marie-Tooth disease who had received a probable SARS-CoV-2 infection, was able to benefit of continuing hand rehabilitation after tendon transfer surgery via tele-rehabilitation. It was also possible to advise the patient on ways to prevent the spread of infection and to cope with restrictions that limited outpatient visits⁵.

In Padua, particular attention was paid to the reorganization of the paediatric palliative care (PPC) referral service. The improvement of their standard of care was achieved by collecting data on the consequences of COVID-19, by holding multidisciplinary meetings to share the approach with professionals involved in PPC in the Veneto region and by preparing educational material on COVID-19 for patients and their families. The data collected were used to assess real needs, and to develop a more appropriate reorganization model⁶. The following strategies were in particular put in place: (i) a 24-hour telephone service assisted by an experienced nurse; (ii) the reduction as much as possible of non-urgent home activities, to favor urgent follow up and critical interventions; (iii) the management of non-urgent needs through advice and 24-hour training for the COVID-19 emergency for healthcare professionals and families; (iv) monitoring and training of patients and families through the use of telemedicine through nursing telephone monitoring

every 15-30 days; (v) converting physiotherapy activity into therapist-led video call interventions; (vi) mandatory SARS-CoV-2 swab testing for patients and relatives prior their admission to the hospitals ⁶.

The PPC referral service in Padua collected also experiences in children with SMA who were obliged to postpone their hospital-based therapy (i.e., infusions of nusinersen®). Forty-eight % of parents of children with SMA perceived a worsening of muscle strength, although no correlation between delayed treatment and changes in functional scores in short and long term assessments was found ⁷. This discordant perception is mainly due to a state of parental anxiety related to the suspension of therapy and physiotherapy, and to a perception of changes in their QoL.

In accordance with this statement, an important study was carried out in Lombardy with the aim to evaluate the consequences of COVID-19 pandemic measures and prolonged home isolation on quality of life (QoL) and perceived disease burden ⁸. Between February and May 2020, 350 NMD patients underwent a telephone interview. The results showed that the virus outbreak impaired some aspects of QoL and affected access to outpatient care and ancillary services, with limited use of remote alternatives ⁸.

At the regional level, a study from the Liguria region analyzed the impact of SARS-CoV-2 infection on patients with NMD and in particular on those affected by Myasthenia Gravis and Guillain Barré syndrome (GBS), in consideration of their standard immunosuppressive therapy. The study concluded that SARS-CoV-2 infection could both cause GBS and affect the outcome of patients with non COVID-19 related GBS ⁹.

An empowerment of national and international networks was done to improve collaboration and encourage the birth of new research projects. Colleagues from the Liguria region participated in the national NeuroCovid project, a multicenter cohort study on neurological disorders associated with COVID-19, conducted in 51 centers in Italy, sponsored by the Italian Society of Neurology (SIN). It seems that a wide spectrum of treatable neurological manifestations may be associated with COVID-19 infection, including hypo-ageusia, hyposmia, acute ischemic stroke, delirium, headache, cognitive impairment, abnormal behavior or psychosis, seizures, GBS, severe encephalopathy with stupor or coma, dizziness, encephalitis and hemorrhagic stroke, and most cases occur in middle-aged adults with mild or severe respiratory syndrome.

On the other side, a national multicenter study under the leadership of tertiary NMD center in Milan documented the disease course and outcome of COVID-19 viral infection in NMD patients, and investigated the po-

tential acute exacerbations of muscle symptoms in these patients. The study concluded that COVID-19 manifestations and morbidity in NMD patients were similar to those presented in the general population and there were no objective changes in disease course during COVID-19 infection.

Central-Southern Italy

The approach of the central-southern regions was different. As part of the reorganization of health- services, NMD experts valued the figure of the family pediatrician as key player in connecting patients and families to health professionals. In particular, the importance of the pediatricians in achieving an early diagnosis was underlined, and in line with a project shared by the Italian Society of pediatricians (SIMPe) and AIM, the training of these figures and their entrustment with the role of “case manager” of the clinical picture, were promoted.

A pediatric epidemiology and research network (RePER) was activated since the outbreak of the pandemic began ^{10,11}, with web-based focus and training on rare conditions NMDs included. RePER created training web pages on various diseases with the aim of improving information, knowledge and care over time. These specific web pages are easy to consult and indicate the characteristics, the warning signs, the diagnostic possibilities, and the specialist reference centers for each disease. For some pathologies, experts have defined a summary of symptoms to consider when raising the suspicion of an infection, useful for implementing the likelihood of a timely diagnosis.

In order to strengthen the network between pediatricians and NMD expert reference centers and to increase knowledge on neuromuscular pathologies, RePER activated a series of web conferences held by experts for training purposes. Finally, the role of pediatricians as “case manager” has allowed a direct relationship with families and integrated commitment with other health professionals, school and specific needs.

Finally, the staff views on the changes in the care provided by a rehabilitation centre as part of a larger project investigating the impact of these changes on professionals, patients and their families were reported ¹². The survey was conducted using an open-ended questionnaire including six-items, on the practical and psychological aspects that emerged during the pandemic, in relation to the healthcare services provided by the centre and to the patients/caregivers conditions. The participants, most of them physiotherapists, highlighted 169 aspects emerging in the pandemic, 48.5% referring to the resources used to cope with critical issues and 51.5% concerning the difficulties encountered. Emotional aspects prevailed on practical aspects both in resources (52.4 vs 47.6%) and

in difficulties (57.5 vs 42.5%) categories. In particular, with regard to patients' resources, psychological benefits, despite the burden, were greater than practical ones (87 vs 13%), in the form of improved intra-family relationships, feeling more cared for, and satisfaction for the received care.

Discussion

Although the experiences reported here, implemented to address the difficulties in maintaining adequate care of the NMD patients at the time of the COVID-19, are different and not integrated in a common national health plan, they summarize few salient and common points as the need for information, the need for training, the need for sharing. During the of COVID-19 outbreak, we saw that the best way to get assistance in NMDs was to strengthen or even create networking by considering all stakeholders.

In fact, only by creating shared paths and with well-organized flows at various levels, it is possible to overcome unexpected events, or insurmountable challenges such what we had to face with rare diseases in general and with NMDs in particular.

One tool that has proved useful in networking is telemedicine that has offered an immediate solution to break down both physical and psychological barriers. The use of telemedicine for visits, rehabilitation and training was the strategy shared by both macro-areas. The telemedicine model was developed not solely because of this need. Previous studies aimed to assess whether this method was adequate and comparable to face-to-face visits. Hosbon et al., in 2016¹³ found that Amyotrophic Lateral Sclerosis (ALS) patients treated with remote approaches had the same level of care and comparable survival as those with face-to-face visits. Furthermore, they reported that a virtual approach seems to reduce emergency room access and acute hospitalizations. Similar conclusions have been reached by Portaro et al.¹⁴ for patients with Facio-Scapulo-Humeral Dystrophy (FSHD).

Another useful tool was the implementation of on-line sharing of critical care and second opinion through videoconferencing consultation. This tool in the age of information technology should be encouraged, by allowing for more frequent meetings, the emergence of new research opportunities and in general, more shared decision-making.

Furthermore, the empowerment of national and international networks has allowed the definition of common guidelines for the management of neurological complications from COVID-19 infection and for vaccinations.

In conclusion, the COVID-19 pandemic has confronted the healthcare system with an unexpected dif-

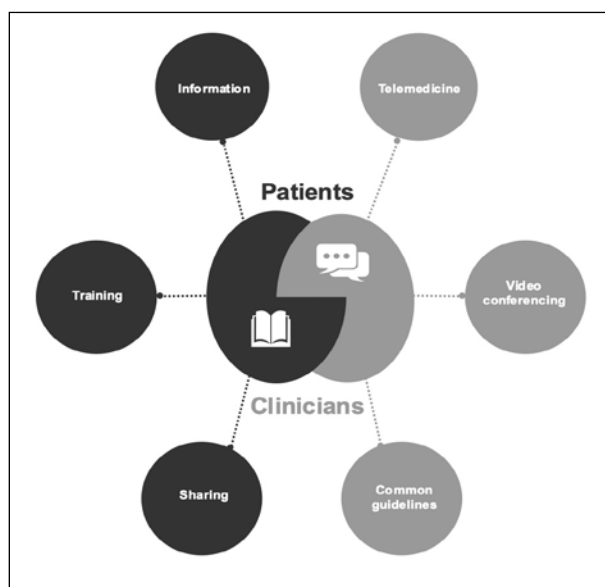


Figure 1. Graphic layout of action strategies implemented to overcome the difficulties imposed by the COVID-19 pandemic.

ficulty in managing patients with rare, debilitating and complex diseases, such as neuromuscular diseases. However, the resilience of the clinicians who deal with these pathologies has made it possible to find and put in action strategies that reduce the distances by favoring connections at multiple levels of intervention (Fig. 1). Of course, the path is still too long, web connections are not always optimal and education to use new tools is not widely spread. However, we are aware that, as a popular saying goes “the road is harder when you’re headed for the sky”.

Ethical consideration

No mention is made of sensitive data referable to patients.

Funding

The Authors did not receive any financial support. The study was supported by the Ministry of Science and Higher Education of Russia, agreement No. 075-15-2021-1346.

Conflict of interest

All the authors declare no conflict of interest.

Author contributions

GA and LP conceptualized this study after having investigated the interest in sharing the information of the

various co-authors. CA, DG, MG and MG_i acquired and described the data concerning their territorial reality.

GM and GA analyzed the available data in the light of the literature and wrote the draft, under the guidance, supervision and methodology of LP. GA and LP reviewed the final version of the paper

References

- ¹ Bertran Recasens B, Rubio MA. Neuromuscular diseases care in the era of COVID-19. *Front Neurol* 2020;11:588929. <https://doi.org/10.3389/fneur.2020.588929>
- ² Natera-de Benito D, Aguilera-Albesa S, Costa-Comellas L, et al. COVID-19 in children with neuromuscular disorders. *J Neurol* 2021;268:3081-3085. <https://doi.org/10.1007/s00415-020-10339-y>
- ³ Mauri E, Abati E, Musumeci O, et al. Estimating the impact of COVID-19 pandemic on services provided by Italian Neuromuscular Centers: an Italian Association of Myology survey of the acute phase. *Acta Myol* 2020;39:57-66. <https://doi.org/10.36185/2532-1900-008>
- ⁴ Solé G, Salort-Campana E, Pereon Y, et al. Guidance for the care of neuromuscular patients during the COVID-19 pandemic outbreak from the French Rare Health Care for Neuromuscular Diseases Network. *Rev Neurol (Paris)* 2020;176:507-515. <https://doi.org/10.1016/j.neurol.2020.04.004>
- ⁵ Prada V, Bellone E, Schenone A, et al. The suspected SARS-Cov-2 infection in a Charcot-Marie-Tooth patient undergoing postsurgical rehabilitation: the value of telerehabilitation for evaluation and continuing treatment. *Int J Rehabil Res* 2020;43:285-286. <https://doi.org/10.1097/MRR.0000000000000418>
- ⁶ Lazzarin P, Avagnina I, Divisic A, et al. Management strategies adopted by a paediatric palliative care network in northern Italy during the COVID-19 pandemic. *Acta Paediatrica* 2020;109:1897-1898. <https://doi.org/10.1111/apa.15411>
- ⁷ Agosto C, Salamon E, Giacomelli L, et al. Effect of the COVID-19 pandemic on children with SMA receiving nusinersen: what is missed and what is gained? *Front Neurol* 2021;12:704928. <https://doi.org/10.3389/fneur.2021.704928>
- ⁸ Gagliardi D, Costamagna G, Abati E, et al. Impact of COVID-19 on the quality of life of patients with neuromuscular disorders in the Lombardy area, Italy. *Muscle Nerve* 2021;64:474-482. <https://doi.org/10.1002/mus.27378>
- ⁹ Garnero M, Del Sette M, Assini A, et al. COVID-19-related and not related Guillain-Barré syndromes share the same management pitfalls during lock down: The experience of Liguria region in Italy. *J Neurol Sci* 2020;418:117114. <https://doi.org/10.1016/j.jns.2020.117114>
- ¹⁰ Doria M, Annicchiarico G, Rachele C, et al. Raccomandazioni per il riconoscimento precoce delle malattie neuromuscolari (Focus sulla Distrofia Muscolare Duchenne).
- ¹¹ Annicchiarico G. Malattie rare: il valore della microrete. Relazione di cura e investimento su una responsabilità che ci gratifica. *Il Medico Pediatra* 2020;29:21-24. <https://doi.org/10.36179/2611-5212-2020-36>
- ¹² Citarelli G, Garofalo C, Esposito MG, et al. Impact of the COVID-19 pandemic on rehabilitation setting. Part 1: professionals' views on the changes in routine care provided by a rehabilitation centre for patients with muscle diseases. *Acta Myol* 2021;40:132-134. <https://doi.org/10.36185/2532-1900-054>
- ¹³ Hobson EV, Baird WO, Cooper CL, et al. Using technology to improve access to specialist care in amyotrophic lateral sclerosis: a systematic review. *Amyotroph Lateral Scler Frontotemporal Degener* 2016;17:313-324. <https://doi.org/10.3109/21678421.2016.1165255>
- ¹⁴ Portaro S, Calabrò RS, Bramanti P, et al. Telemedicine for facio-scapulo-humeral muscular dystrophy: a multidisciplinary approach to improve quality of life and reduce hospitalization rate? *Disabil Health J* 2018;11:306-309. <https://doi.org/10.1016/j.dhjo.2017.09.003>