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Healthcare reassessment in a pandemics time: challenges for CF



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Northern Italy has been the first sizeable European region largely affected by COVID-19. At the time being (March 15th 2020) the Italian Ministry of Health reports 20,603 SARS-CoV-2 positive cases and 1,809 deaths [1]. This is an opportunity of witnessing how COVID-19 is affecting people with Cystic Fibrosis (pwCF) not just in terms of clinical expression, but also by the reshaping of social behaviours, access to specific therapeutics, and models of care.

In Italy COVID-19 has been reported (so far) in two mildly symptomatic CF patients, one adult and a newborn child, whose clinical evolution is still unclear (personal communications). The diffusion of the virus in the general population and the increasing number of severely ill people have understandably been a substantial cause of concern for the national CF community, the major worry being that the CF patient is more vulnerable to a severe clinical course of COVID-19. Although this is not yet known to be true, the 2009-2010 pandemic Influenza A (H1N1) could perhaps offer some insights. Patients with CF infected with H1N1, particularly those with advanced stage lung disease, showed increased morbidity and a higher case fatality rate compared to healthy controls as well as patients with other chronic respiratory diseases [2,3]. COVID-19 is a different respiratory disease, but it seems reasonable to assume that those with a pre-existing lung condition, like CF, would be at greater risk of more severe manifestations of disease than are healthy people. Several Italian CF centers are joining in a network that will collect COVID-19 cases and will hopefully provide the data so urgently needed.

In an epidemic context, complicated by the present uncertainties and quite volatile circumstances, it is extremely challenging to convey conclusive messages. The Italian government decided to seal off two large regions where there were foci of infection. This was communicated on the February 22-23 weekend with great emphasis with 24 hour coverage in all of the media. Just a few days later, regional authorities and several media challenged these provisions, voicing concerns about the everyday life and long-term economic consequences. The harsh public debate that followed weakened the determination of the population to adhere to the infection control procedures. Words of caution from the scientific community were not given proper attention by many people in the community, underestimating the seriousness of the situation, which in turn reduced the efficacy of the prevention measures. Two weeks later the rapidly increasing trend of infections led the government to issuing even more draconian measures involving

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the whole nation. These inconsistencies confused and upset the population, but were even more disturbing for pwCF, who are well aware of the potential personal impact.

A further cause of concern was the message continuously repeated by the media that COVID-19 deaths concerned mainly elderly people or those with concomitant diseases. Although this was meant to reassure the general population, it had the opposite effect on those suffering from chronic diseases, including pwCF. The news reporting shortage of ICU beds and advanced care technology made some pwCF worry about reduced access to lifesaving therapies should they suffer severe pulmonary exacerbations.

Hospital regulations changed repeatedly while the SARS-CoV-2 story was evolving, becoming progressively stricter but also more difficult to interpret for those with chronic pulmonary diseases like CF. Initially, some health institutions phoned patients before scheduled admissions, aimed at exploring if patients had visited high risk zones or had been in contact with a suspect or confirmed case of SARS-CoV-2 infection during the previous fortnight. The interview, which included questions on the presence of fever, dyspnea or cough, was meant to single out individuals at risk of infecting others, and these patients were asked not to go to the hospital. This pre-clinic triage had evident limitations when used in pwCF, who have daily respiratory symptoms; it may make it difficult to distinguish classic CF pulmonary exacerbations from the first signs of COVID-19, and hence risk unnecessarily delayed appointments. Eventually, as SARS-CoV-2 cases increased, most hospitals stopped non-urgent activities and limited access to wards and clinics to urgent cases only. Because of the impossibility to predict the end of the critical phase, pwCF could not be immediately rescheduled, causing disruptions to the essential monitoring of patients defined in our standards of care [4].

The limited access to hospital services highlights the significance and urgency of alternative methods of care, such as telemedicine [5]. CF is a chronic condition best served by regular monitoring by a multidisciplinary team; much of this care is ideal for telehealth and there have already been positive experiences reported [6,7,8]. In Italy, a few pilot programs have been implemented [9,10], but telemedicine is far from routine use and there are hurdles to its practice in today's critical context. Patients and families must have access to the internet. While many team members are able to establish sufficient conversation, there are other aspects of care that require a face-to-face interaction (e.g. performance of hands-on pulmonary therapy). The need for spirometry, laboratory monitoring, and sputum cultures might be circum-

vented by home technology and other local resources, but they are not universally reliable.

COVID-19 is also having a significant impact on CF research in Italy. Many investigators are currently prohibited from their work in order to limit the risk of cross-infections and viral dissemination. Travel limitations and staff shortages are making appointments scheduled for clinical studies much harder to keep. Offers of rescheduling are not infrequently declined by trial participants concerned about exposure, with a serious risk of loss of data and problematic drug distribution.

All of these issues point to a CF disadvantage in the current pandemic crisis. This is only partially balanced by the familiarity of pwCF with infection control measures, such as the use of face masks, advice to avoid contacts with other CF patients and segregation policies adopted in CF Centers. They are probably better equipped for a new way of social life than healthy individuals, who are now paradoxically faced with regulations and procedures they had previously encountered only in fictional situations, like in the "Five Feet Apart" movie [11]. While this may offer a theoretical advantage in the current context, we should not jeopardize it with inconsistent communication and limited consideration of CF peculiarities.

COVID-19 has modified patients' and health professionals' lives and ways to handle CF. It is vital to keep pwCF and their families informed, adapt to a situation which is not only unprecedented but also changing by the day, and continue to invest in research because, to quote what I was recently told, "eventually the virus will go, CF will stay". That is undoubtable, but it is also likely that at the end of the pandemic, the way we deal with CF will be different. Some changes may be positive, like an acceleration in the use

of telehealth technologies and remote monitoring of clinical status, for others, like a possible shift in research directions or different choices in healthcare investments, it is hard to foresee the impact on CF

References

- http://www.protezionecivile.gov.it/media-communication/press-release/detail/-/asset_publisher/default/content/coronavirus-sono-20-603-i-positivi
- [2] Renk H, Regamey N, Hartl D. Influenza A(H1N1)pdm09 and cystic fibrosis lung disease: a systematic meta-analysis. PLoS One 2014;9:e78583.
- [3] Viviani L, Assael BM, Kerem E. ECFS (A) H1N1 study group. Impact of the A (H1N1) pandemic influenza (season 2009-2010) on patients with cystic fibrosis. | Cyst Fibros 2011;10:370-6.
- [4] Conway S, Balfour-Lynn IM, De Rijcke K, Drevinek P, Foweraker J, Havermans T, et al. European Cystic Fibrosis Society Standards of Care: Framework for the Cystic Fibrosis Centre. J Cyst Fibros 2014(Suppl 1):S3–22 May.
- [5] Hollander JE, Carr BG. Virtually Perfect? Telemedicine for Covid-19. NEJM 2020:382:1679–81.
- [6] Bell SC, Mall MA, Gutierrez H, Macek M, Madge S, Davies JC, et al. The future of cystic fibrosis care: a global perspective. Lancet Respir Med 2020;8:65–124.
- [7] Choyce J, Shaw KL, Sitch AJ, Mistry H, Whitehouse JL, Nash EF. A prospective pilot study of home monitoring in adults with cystic fibrosis (HOME-CF): protocol for a randomised controlled trial. BMC Pulm Med 2017;17:22.
- [8] Wood J, Jenkins S, Putrino D, Mulrennan S, Morey S, Cecins N, Hill K. A smart-phone application for reporting symptoms in adults with cystic fibrosis improves the detection of exacerbations: Results of a randomised controlled trial. J Cyst Fibros 2019 Sep 12. pii: S1569-1993(19)30887-2.
- [9] Grzincich G, Gagliardini R, Bossi A, Bella S, Cimino G, Cirilli N, et al. Evaluation of a home telemonitoring service for adult patients with cystic fibrosis: a pilot study. J Telemedicine Telecare 2010;16:359–62.
- [10] Tagliente I, Trieste L, Solvoli T, Murgia F, Bella S. Telemonitoring in Cystic Fibrosis: a 4-year Assessment and Simulation for the Next 6 Years. Interact J Med Res 2016;5:e11.
- [11] https://en.wikipedia.org/wiki/Five_Feet_Apart