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Impact of COVID-19 on people with cystic fibrosis

Early and detailed characterisation of coronavirus disease 2019 (COVID-19) has emerged principally through publications from China, where the disease was first identified.¹ As the pandemic spread to Lombardia, Northern Italy and then globally,² evaluating the impact on people with cystic fibrosis has become imperative, because the prevalence of this inherited condition is much higher in populations derived from Europe than in other populations.³ Since the beginning of the outbreak in Lombardia, severe acute respiratory syndrome coronavirus 2 (SARS-CoV-2) infections have been reported in ten patients with cystic fibrosis out of 42 161 people in the region known to have been infected. The number of infected patients in Lombardia make up a substantial proportion of the 101 739 total SARS-CoV-2 infections in Italy (according to available data on March 31, 2020). The Italian patients with cystic fibrosis were resident in endemic areas and acquired the infection from family members. These observations, as well as data from other European countries (five patients with cystic fibrosis in France have been reported to have SARS-CoV-2 infection, seven in the UK, five in Germany and three [one transplanted] in Spain) suggest few patients with cystic fibrosis, mainly adults, are becoming infected with SARS-CoV-2, without apparent effect on cystic fibrosis disease severity. From these data, it is not possible to identify factors that might be protective, for example use of long-term antibiotic therapy such as azithromycin.

The few reported cases of SARS-CoV-2 infection in people with cystic fibrosis might reflect the efforts of families across Europe to minimise social contacts. This effort might be considered a success, but

there is no room for complacency and the directive for social shielding of both the patient and family members remains clear and important.

More data should be collected to better characterise the impact of COVID-19 on patients with cystic fibrosis. The European Cystic Fibrosis Society (ECFS) is supporting the collection and timely sharing of data from across Europe. The ECFS Patient Registry, in cooperation with national cystic fibrosis registries, has established a data collection and reporting system, contributing to an international harmonised dataset, to identify factors that predict severity of COVID-19.

Viral respiratory tract infections are more severe in patients with cystic fibrosis than in the general population, with an increased risk of complications and a negative impact on lung function. During the 2009 influenza pandemic, H1N1 virus caused substantial morbidity in patients with cystic fibrosis, and in a subgroup with severe lung disease, H1N1 infection was associated with respiratory deterioration, mechanical ventilation, and even death.^{4,5}

The situation in Italy has been monitored carefully to inform management strategies and provide clear advice for patients with cystic fibrosis and their families. Consistent with the Italian Government, the Cystic Fibrosis Center in Milan promptly recommended self-isolation for these patients. Recommendations for preventive measures that were already established in this population, such as using face masks and practising adequate hand hygiene, were reinforced. The cystic fibrosis team in Milan cancelled routine clinic appointments to prevent unnecessary hospital visits and viral spread. Procedures such as respiratory function testing and bronchoscopy were put on hold. Phone calls and email contacts were used to monitor the clinical condition of patients and to provide psychological support immediately after the lockdown.

These measures have subsequently been adopted by cystic fibrosis centres across Europe. There is no doubt that cancelling routine cystic fibrosis clinics and monitoring according to ECFS standards of care³ will have a negative impact on the wellbeing of patients with cystic fibrosis over time. It is important that patients and their families are provided with tools to support self-monitoring during this period. This self-monitoring might include transmission to clinicians of spirometry and oxygen saturation data that are recorded at home. Support should also include the capacity to carry out respiratory culture at home and send to the laboratory securely.

Home visits by health-care professionals present a risk of SARS-CoV-2 transmission and virtual clinics should be used to provide families with advice on all aspects of cystic fibrosis care management, including airway clearance and maintaining exercise. In case cystic fibrosis team members are seconded to work on the front line of tackling this pandemic, it is imperative that lines of communication remain open and that patients with cystic fibrosis do not feel isolated. For patients with more severe cystic fibrosis, access to lung transplantation might be compromised.

People with cystic fibrosis have a phenotypic spectrum ranging from excellent respiratory health to chronic airways disease with productive cough and respiratory compromise. The clinical features of COVID-19 (dry cough, myalgia, and fever) are quite distinct from the symptoms of cystic fibrosis. Therefore, most COVID-19 in people with cystic fibrosis should be recognisable, but it is possible that mild disease might be labelled as within the normal spectrum of symptoms for that individual. A low threshold for testing is therefore needed in this population.

Many isolated families are concerned about collection of medicines and food during this pandemic. Most countries have established systems to ensure



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For information from Italy on COVID-19 see <https://www.salute.gov.it/nuovocoronavirus>

deliveries to people who are isolated, often using volunteers, but as the pandemic progresses, maintaining these systems will be a priority for patients with cystic fibrosis and their families. A further concern is the halt to the development of new therapies for cystic fibrosis.³ No new clinical trials are being initiated in Europe and recruitment of patients to ongoing trials is being stopped. Patients in existing trials are being supported by reducing study visits with the agreement of regulatory bodies such as the European Medicines Agency and by providing study medication to the home.⁶

As national advice changes rapidly to respond to the pandemic, it is important that cystic fibrosis organisations place this advice in context and present information for patients and their families that is based on clear evidence and not conjecture.

This pandemic is imposing an extraordinary level of stress on health services and the consequences include the trauma of witnessing so many deaths, even of young and healthy people, and the impact on the whole community of fear, isolation, and uncertainty for the future. Following the situation in Italy, a community-oriented plan for the next pandemic is clearly needed.⁷

People with cystic fibrosis and their families have invested considerable time and energy to maintain good health and, now, on the cusp of remarkable new therapies to transform their condition, they face a global pandemic, the effect of which is unclear. Early data suggest that most patients with cystic fibrosis are doing an exceptional job avoiding SARS-CoV-19 infection, but they must remain dedicated to this task, as data are gathered from across Europe to better understand factors that affect the severity of COVID-19 in people with cystic fibrosis.

We declare no competing interests.

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