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COVID-19 and cystic fibrosis. Background and recommendations

COVID-19 y fibrosis quística. Antecedentes y recomendaciones

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Mr. Editor:

The cystic fibrosis is a multisystemic genetic entity, the prognosis of which depends on early diagnosis and interdisciplinary therapeutic management (1). It presents an autosomal recessive pattern of inheritance, caused by mutations in the cystic fibrosis transmembrane conductance regulator gene (CFTR) located at 7q31.2, which encodes an ion channel protein, with more than 2,000 identified mutations (1, 2). It is the most common lethality risk disorder in the United States, affecting approximately 1 in every 4,000 newborns. However, in some European countries it has a higher incidence (1). Estimates suggest that more than 72,000 people live with cystic fibrosis worldwide (2).

In Chile, with the existing ethnic mix, an approximate incidence of 1 in 8,000-10,000 live newborns is estimated, which means approximately 25-30 new cases of cystic fibrosis per year (3). In turn, it is covered by the Explicit Health Guarantees (number 51, specifically), which allows access, quality, opportunity and financial protection. According to the data offered by this system, there is an estimated demand of 573 people with this entity in the country, for the year 2017 (4).

In the face of the 2019 coronavirus pandemic (COVID-19), renamed as Severe Acute Respiratory Syndrome Coronavirus 2 (SARS-CoV-2), initial data suggests that exceptional work is being done to prevent the infection in individuals with cystic fibrosis (5), and further collaborative studies will be required to better understand the factors that affect the severity of COVID-19 in people with this genetic disorder (5,6).

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At least ten patients with cystic fibrosis and COVID-19 infection have been documented in Lombardy, region of Italy, according to data collected at the end of March 2020. Similarly, seven cases have been described in the United Kingdom, five in France and Germany, each one, and three in Spain, one of which was transplanted (5). The first case of infection occurred in an Italian adult with cystic fibrosis, who presented mild symptoms (6). Similar evolution presented other adult patients, who did not show an apparent effect on the severity of the disorder. However, it is not possible to identify protective data with these data, such as the use of long-term antibiotics, such as azithromycin. The low frequency of infection may reflect the efforts of the patient and their family members to minimize contacts (5), an important aspect that must be promoted at all levels.

Likewise, the first pediatric case was documented in Italy in a one-month-old boy, with positive detection for cystic fibrosis in the neonatal investigation and confirmed with a molecular study. Presents as a positive family history, a paternal cousin affected with the same genetic entity. Its management is carried out with nebulization of saline solution, pancreatic enzymes and fat-soluble vitamins, without isolating microorganisms in the last nasopharyngeal aspirate. The patient never developed a fever, or any other sign of infection. The contact was through his grandfather who presented asymptomatic infection for COVID-19 diagnosed by real-time PCR (6).

Based on the limited available evidence, the benign clinical course in the pediatric population remains unknown. It is hypothesized that this may be due to the high plasticity of your immune system, low expression of ACE2 receptors or the exposure of other coronaviruses that may be common in this period (5). Therefore, more data should be collected to better characterize the impact of COVID-19 in cystic fibrosis patients regardless of age.

In fact, the organizations and different groups of patient registries have established a reporting

system, which contributes to a data set uniformly and internationally, to better identify the factors that predict the severity of COVID-19. These data will be carefully monitored to inform management strategies and provide advice for patients and their families (5).

Recommendations

As previously mentioned, the isolation of the patient and his family should be carried out (5,6), as well as reinforcing preventive measures, such as the use of face masks and proper hand washing. The suspension of routine clinical appointments in the hospital is convenient, as well as the performance of nebulisations, respiratory function tests and bronchoscopies to avoid contagion in health centers. The Telemedicine can be used for this, and psychological support can also be provided through this medium. These measures were taken by the Center for Cystic Fibrosis in Milan, and followed throughout Europe. The patients and their families should receive the necessary tools for self-monitoring during this period, which include the transmission of the results to the doctors of oxygen saturation, spirometry and the ability to perform respiratory culture at home and send it to the laboratory in a safe mode. Furthermore, it is a priority to guarantee the delivery of medicines and food to isolated people and their families, while the pandemic lasts (5).

Similarly, the home visits by health professionals can represent a risk of transmission, so virtual clinics must provide advice on all aspects of care (5). These should include the basics of respiratory kinesitherapy, properly perform to the nebulisations, clearing of the airways, and exercise regularly. In the event that a family member must leave the home, either for work or for another strictly necessary cause, the necessary hygiene measures must be provided to avoid the transmission of the virus.

Finally, there is concern about the interruption of adequate control in patient care centers, as well as the development of new therapies as they cannot start clinical trials (5). Although some Latin American nations are carrying out similar protection activities, actions such as those undertaken in Europe can be replicated in the region and, if results are obtained, they can be compared and conclusions can be drawn according to the characteristics of the population.

In memoriam of the Professor Nancy Freitez de Sardi, who was Senior Lecturer in the Public Health course of the Department of Preventive and Social Medicine of the Faculty of Medicine of the University of Los Andes in Mérida, Venezuela. She was a teacher and a friend, an academic and an ecologist who left a legacy in her students, in the city of eternal snows and in the country.

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